Comments to ear embryology

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External ear

Auricle is forming from 4.week form 6 paired prominences of mesenchymal tissue of first mandible and second hyoid brachial arch.

From mandible arch grow:

- **Tuberculum tragicum**
- **Tuberculum helicis**
- **Tuberculum helicis intermedium**

From hyoid arch grow:

- **Tuberculum anthelicis**
- **Tuberculum antitragicum**
- **Tuberculum lobulare**

Auricle is formed by merging those tuberculi until 3.month. Ectoderm of first branchial sulcus forms concha auriculare. Auricle is primary located caudally, but with mandible evolving (until 20.week) it migrates cranially to its common location. In 4-5 ears old has auricle 80% of its size; in 9 years has its common size.

External auditory canal evolves form ectoderm of first branchial sulcus. Ectoderm forms strip from 4.week, which migrates medially to meet endoderm from first branchial fissure. This strip canalizates from 28.week. Cartilaginous part of auditory canal corresponds with primary auditory canal, bone part corresponds with epithelial strip. Fibrous layer stays between auditory canal and tympanic cavity, which is base of eardrum. Medially from eardrum is endoderm of first branchial exagination, from which is formed middle ear cavity. Ossification of bone part of auditory canal starts in 12.week.

Middle ear and Eustachian tube (ET)

Middle ear and ET evolve form tubotympanal recessus formed by 1.entodermal pharyngeal pocket, which appears in 3.embryonic week. This pocket contracts in 2.month into bottle shape. Contracted part lengthens and forms ET. Blind external end gets wider and is divided into 4 pouches: anterior, middle, posterior and superior, which form tympanic cavity and pneumatization of temporal bone. Middle ear cavity is filled with mucous mesenchymal tissue during its evolving. From 3.embryonic month this mesenchymal tissue dissolves and starts resorpting. Complete resorption last until 1.year of age and sometimes even longer.

Musculus tensor tympani is formed from mesoderm of first branchial arch, innervated is by mandible branch of n.V. Musculus stapedius evolves from mesoderm of 2.arch, innervated is from n.VII.

Malleus and incus evolve from 4.week from Meckel´s cartilage of 1.arch (neck, head of malleus, short processus and body of incus) and from Reichert´s cartilage of 2.arch (manubrium mallei, long processus of incus and structures of stapes). Medial part of stapedic plate and ligamentum anulare stapedis evolve from otopocket. Ossification of incus and malleus starts in 15.week and in 24.week is complete. Stapes evolvement starts in 4.week as a stapedic circle around arteria stapedia. Ossificates between 18-24 week.

Antrum mastoideum

Evolves as an extension of epitympanum in 21.-22.week. Pyramid pneumatization starts from 28.week, pneumatization of mastoids starts from 33.week. Pneumatization of temporal bone is complete years after birth and this process can be ended even in adults.

Interior of ET is forming from persistent first pharyngeal evagination. Entodermal part of evagination extends laterally and its distal part extends towards middle ear. Musculus levator veli palatinin and tensor veli are formed in 10-12.week. ET grows and its lumen is starting to evolve, in 10.week has 1mm, in birth has 13mm. Most significant growth proceeds in cartilaginous part. Skull base is flat in neonate, so that ET is in early childhood practically horizontal.

Nervus VII. and VIII.

N.VII. is nerve of 2.branchial arch. It can be identified from 3.week together with statoacoustic nerve as a cell aggregate – acousticofacial ganglium – ventrally from placoda oticum. They can be differentiated in the end of 4.week. N.facialis is ventrally on upper surface of 2.branchial arch. Motoric part of n.VII evolves separately from neuroblasts in upper part of rhombencephalon in vicinity of pons Varoli (like n.VI) which can clarify damage of both nerves in Moebius´s syndrome. Sensoric n.intermedius comes from ganglion geniculate from 7.week. Abnormalities often afflict angle in 2.knee of nerve during its course in middle ear wall (60 grades compared to normal 120 grades), which moves nerve between round and oval window.

Nervus VIII. Ganglion acusticofaciale n.VIII has common base with ggl.geniculi n.VII, later separates. Ggl.n.VIII is placed to medial part of auditory pouch wall and divides itself into 2 parts: upper for ggl.vestibulare, lower for ggl.cochleare.

Inner ear

Inner ear is evolving in 3.-4.week on lateral part of head as an ectodermal thickening called acoustic placod. This structure gets deeper and forms socket, its opening is closing and forms so called otocyst coated with ectoderm and surrounded by mesechyme. Surrounding mesenchyme starts to formcartilaginous capsule of otocyst, which ossificates around 17.week. During 5.week otocyst differentiate by 3 plicae into dorsal vestibular part (utriculus, semicircular ducts and ductus endolymphaticus) and ventral cochlear part (sacculus and ductus cochlearis). Membranous labyrinth is formed during 6.month. Otocyst probably forms cells of n.VIII. Ggl.n.VIII is divided into upper (for utriculus, lateral and superior semicircular canals) and lower part (for sacculus, posterior semicircular canal and cochlea).

Clinical anatomy of ear

Ear is peripheral auditory and balance analyzer. The ear consists of free major parts: the external ear, the middle ear and the inner ear. The external ear includes an auricle, the external auditory canal and the tympanum. The middle ear includes pneumatic system of mastoid process, which is connected via aditus ad antrum with cavum tympani. The eustachian tube connects the cavum tympani with the cavity of the middle ear. The inner ear is composed of bone and membranous labyrinth.

Content

Auricle

The auricle is formed from elastic cartilage covered with skin. Perichondrium tightly adheres to the skin at ventral side. Perichondrium and skin are dorsally connected with thin layer of connective tissue. Earlobe is formed from fat tissue. Angle between the auricle and skull shouldn´t be more then 15 degrees. On auricle we can differentiate following structures: helix, anthelix and between them fold (scapha).
Anthelix is ventrally divided into crura anthelicia; between them is located fossa triangularis. Concha auriculae is divided by crus helicis into cymba conchae and cavum conchae. Entrance to external auditory canal is surrounded by tragus and antitragus, they are divided by incisura intertragica. Incisura heliotragica is located between helix and tragus – place of incision in endaural approach to middle ear. Form of external ear allows sound concentration from outside into auditory canal. Auricle is innerved from n.V, VII, IX and from 2.and 3.cervical nerve (via n.auricularis magnus and n.occipitalis minor)

External auditory canal (meatus acusticus externus)

External auditory canal starts in auricle, goes through temporal bone and is ended by eardrum. Outer third of auditory canal is formed by skin, which is connected with cartilage by thin layer of fibrous tissue. Skin in this area contains hair follicles and glandulae ceruminosae et sebaceae.
Gl.ceruminosae are modified apocrine glands producing secretion, which gets brown on air. This secretion together with sebaceous glands secretion form cerumen. IT contains triglycerides and fat esters as well. Medial two thirds of auditory canal are formed by bone with skin of canal. Auditory canal is innerved by n.VII and IX.

Eardrum (membrana tympani)

Eardrum in adults has size 9x10 mm.

- **Pars tensa** is formed by 4 layers. From external auditory canal towards middle ear: epithelium, radial and longitudinal fibrous layer and mucosa. Eardrum is connected with auditory canal wall by fibrous annulus fibrocartilagineus.
- **Pars flaccida** (Schrapnell´s membrane) is located above mallear prominence between stria mallearis anterior and posterior. Pars flaccida is more thin than pars tensa, it doesn´t have longitudinal fibrous layer.

Cavum tympani

- **Epitympanum** (atticus) is area between upper part of eardrum and tegmen tympani of temporal bone.
- **Hypotympanum** is circumscripted by lower edge of eardrum and base of eardrum cavity
- **Mesotympanum** is part of tympanic cavity medially from eardrum
- **Protympanum** lies ventrally from eardrum and there is orifice of ET
- **Prussak´s space** is circumscribed by pars flaccid, ligamentum mallei lateralis, neck of malleus and dorsally is opened to epitympanum. IT is the most frequent place of retraction pocket and cholesteatoma formation.

ET and ventral part of middle ear cavity is lined with columnar epithelium with cillia, which merge into cilliary epithelium of nasal cavity and nasopharynx. Towards mastoid process we can see that epithelium is getting low and flat and number of cilia is decreasing. Dorsal part of middle ear cavity is lined with one-layer cubic epithelium and in mastoideal pneumatization we find flat epithelium without cilia.

Middle ear ossicles, muscles and nerves

Middle ear ossicles connect eardrum and oval window

- **Malleus**: is connected with medial surface of eardrum. Processus brevis and inferior part of manubrium (umbo membranae tympani) are tightly connected with fibrous layer of eardrum. Middle part of manubrium is connected with mucosal layer – stria mallearis. Neck and head are located in epitympanum. Manubrium mallei is connected with incus.
- **Incus:** processus brevis goes dorsally, processus longus is connected with stapes by processus lenticularis
- **Stapes**: head of stapes is connected with plate by two branches, anterior and posterior. Plate is held in oval window by ligamentum annulare stapedis. Plate has thickness about 1,5-3 mm
- **M.tensor tympani** is located in temporal bone paralelly with ET. After entering middle ear its tendon goes towards processus cochleariformis, is turned around it and is connected to medial surface of neck and head of malleus
- **M.stapedius** leaves eminentia pyramidalis on dorsal wall of middle ear cavity. Tendon is connected to head and posterior branch of stapes.
- **N.VII (n.facialis)** goes out of pons and enters upper frontal part of internal auditory canal, where is joined together with n.intermedius, which brings olfaction fibers. It goes through **Fallopi´s canal**, comes out from temporal bone in formen stylomastoideum and goes along dorsal part of m.digastricus. Its main stem enters glandula parotis, where is divided into 6 branches for mimic muscles

In temporal bone are derived from n.VII:

- **N.petrosus maior** brings parasympathetic fibers to ggl.pterygopalatinum and innerves lacrimal glands and glands in mucosa of palate and nasal cavity. It goes towards canalis pterygoidei, where is joined with sympathetic n.petrosus profundus and forms canalis pterygoidei
- **N.stapedius** innerves motorically m.stapedius
- **Chorda tympani** goes in middle ear between manubrium mallei and processus longus incudis. IT goes out in fissure petrotympanica. It connects with n.lingualis and leads

parasympathetic fibers into ggl.submandibulare (for submandibular and sublingual salivary glands) and sensoric olfactionfibers (for anterior 2/3 of tongue)Recessus facialis is V-shaped area between chorda tympani and n.VII

- **Arnold´s nerve (r.auricularis n.X)** is branch of n.X, which contains also fibers from n.IX and goes above vault of jugular bulbus. Upper fibers ae connecting with n.facialis, lower ones lead sensitive information from posterior surface of external auditory canal. Arnold´s reflex can be induced by irritation of posterior wall of external auditory canal.
- **N. tympanicus (Jacobson´s nerve)** is branch of n.IX. It forms plexus in middle ear. N.petrosus minor goes out of this plexus. It goes from brain cavity through fissura petrotympanica and leads parasympathetic fibers into ggl.oticum. From this ganglion is parasympathetically innerved specific salivary gland by n.auriculotemporalis.

Eustachian tube (ET)

ET is connection between middle ear and nasopharynx. ET has 3 parts – cartilaginous, junction and bone. Anteromedial part is cartilaginous and opens into nasopharynx in the area of torus tubarius, posterolaterl part is bone and opens into middle ear. Junction is located between two previous parts and was called isthmus. Dilatation of cartilaginous part is maintained in children only by m.tensor veli palatine with its medial fibers (m.dilator tubae), which is innerved from n.V (trigeminus). Children with malfunction of this muscle (e.g. with palatal fissures) have dysfunction of ET. Adults have in addition m.levator veli palatine.

ET is rapidly lengthening in early childhood and in 7 years old children has same length as in adults. Neonates have ET shorter by half in comparison with adults (18 mm : 35 mm). Short ET can be accused of protective function disorder.

Young children have ET horizontally placed with maximum deviation of 10°. In adults is this deviation 45°.

Physiological functions of middle ear and Eustachian tube

Middle ear functions

- Sound is transmitted with help of ossicles from 52mm eardrum surface to 3mm surface of oval window. Sound energy is amplified this way to 17:1. In addition, manubrium mallei is 1,3 times larger than long processus of incus, so another 1,3 times amplification of sound energy occurs. Total amplification is 22:1 which means 25 dB.

- There is diffuse gradient between atmospheric pressure and mucosal circulation. Middle ear mucosa can continuously absorb gases, which permanently lower middle ear pressure

ET functions

- **Ventilation** function of ET is maintained by periodic opening of ET. It helps to pressure equalization between middle ear and nasopharynx (middle ear and atmospheric pressure)

- **Drainage** function of ET is maintained by cilliary epithelium in ET and anterior part of middle ear (mucocilliary clearance) and by muscle functions (muscular clearance). Cilliary cells drain secretion into nasopharynx, formed by calyciformis cells or formed during inflammations.

- **Protective** function of ET is maintained by shape and course of ET, muscle activity of soft palate, immunologic and mucocilliary function of mucosa. It lowers risk of infection penetration into middle ear during inflammations of respiratory airways.

ET function disorders

- Closed ET – opens less often than usually (in average ones in 2 minutes in healthy people) according to pressure gradient between nasopharynx and middle ear

- Open ET (patulous, semipatulous) – opens more often than usually or is permanently patent

Pneumatic system of temporal bone

Mastoid process is connected with middle ear through aditus ad antrum. In adults is inside extensive pneumatization. Only small central cavityis developed after birth – antrum mastoideum, which is the base for other cavities. Development and extent of mastoid process pneumatization depends on genetic factors and on number and courses of middle ear inflammations. Children have small mastoid process, pneumatization is low. This small air volume can be cause of easily developing low pressure in middle ear in children. Pneumatization of most parts of temporal bone is complete in 5.-10.years.

Bone labyrinth

Includes bone structures of pars petrosa ossis temporalis (vestibulum, cochlea and semicircular canals)

- **Bone cochlea**: lies in anterior part of bone labyrinth and is formed by spiral bone canal (ductus cochlearis), which whips around 5mm long bone axis (modiolus). Modiolus goes meatusrocaudally from anterior part of upper wall of inner auditory canal. Cochlea has 2,5 screws and is 31-33 mm long. Basal cochlear screw looks in middle ear as a promontorium. Bone plate (lamina spiralis ossea) goes from modiolus lengthwise cochlea, on it are connected basillary and Reisner´s membrane. Bone cochlea is divided by these on 3 spaces (scala tympani, scala media, scala vestibuli). Between bone and membranous cochlea is perilympha of similar composition as a cerebrospinal fluid (with high sodium and low kalium). Inside membranous labyrinth is endolymph, which has high kalium and low sodium (as a intracellular fluid)

Membranous cochlea

- Is located inside bone cochlea

Basillary membrane: is bound to lamina spiralis ossea and divides scala media and scala vestibuli. Externally located scala vestibuli connects with oval window. Cortiś organ is located on basillary membrane in scala media

Reisner´s membrane: is bound to lamina spiralis ossea and divides scala media and scala tympani. Scala tympani is connected with round window. Scala tympani and vestibuli are connected with helicotrema (scala communis) on the apex.

Membrana tectoria: goes from ligamentum spirale, which is bound to lamina spiralis ossea.

Sound is transmitted from stapedial plate via oval window into perilymph in scala vestibuli. Waves in perilymph cause irritation of frequency specific parts of basillary membrane (lower frequencies on the base, higher on the apex)

- **Utriculus and sacculus:** are vesicles containing neuroepithelial receptor cells in the macula sacculi et utriculi. These areas of sensoric epithelium produce gelatinous substance forming otolith membrane. This substance contains crystals of calcium carbonate (otoconia). Macula sacculi is located in ventral plane on medial wall of sacculus, macula utriculi is located on anterolateral wall of utriculi perpendicularly to macula sacculi. Receptors in maculi contains ciliary cells, their cilia goes into otolith membrane with otoconia. Cilliary cells are surrounded by auxiliary cells. Utriculus is ovoid and is sensitive to linear acceleration. Sacculus is smaller than utriculus, is spherical and is connected with cochlea via ductus reuniens.
- **Semicircular canals:** are three – upper, lateral and posterior. They have neuroepithelial cells in ampullar endings connected with utriculus. Upper and posterior canals are on the other ends connected into common opening, which is located in middle part of utriculus. Receptor cells in ampullar endings are formed by cilliary cells. Their cilia are plunged in gelatinous substance form polysaccharides and keratin. Semicircular canals are sensitive to angular acceleration.
- **Ductus and saccus endolymphaticus**: their main function is endolymph absorption and pressure equalization between cerebrospinal fluid and endolymphatic system.

Examination methods of ear

The include sight (including otoscopy) and palpation, imaging methods and functional examinations. Hearing examinations are divided according to patient´s compliance into subjective (patient responds during examination) or objective (patient´s respond is not needed, information are gained by doctor or machine)

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6. Imaging methods

Examination of Eustachian tube function

- **Tympanometry**
- **Valsalv´s maneuver:** expiration against closed nasal entrance increases pressure in nasopharynx and causes opening of ET and pressure increase in middle ear
- **Politzeration**: pressure increase in nasal cavity and nasopharynx with help of ball inserted into nostril with contemporary closure of other nostril. It leads to ET opening and pressure increase in middle ear. Patient contemporary says syllables, which causes velopharyngeal closure (in czech: kuku, káva). Opening of ET and air passing into middle ear cause murmur, which can be registered by otophone (tube connecting ear of examined person and doctor). Patient indicates change of hearing
- **Catheterization**: with help of metal catheters ET opening can be sounded and with ball we can increase pressure in middle ear (similarly like in politzeration). Examination is unilateral and anaesthesia is required (local or general).
- **Toynbee test:** swallowing in closed nasal entrance and mouth leads at first to positive pressure in nose and nasopharynx (1.phase), after that to pressure decrease (2.phase). In 1.phase can air flow into middle ear and cause overpressure. During 2.phase is in middle ear underpressure or overpressure persisted from 1.phase. In case of ET malfunction it is not opened and pressure in middle ear stays same. If eardrum isn´t damaged, test can be evaluated by tympanometry. If pressure in middle ear decreases, function of ET is probably normal. If pressure doesn´t decrease it doesn´t mean malfunction of ET, but other test must be made.
- **Experimental examinations of ET function**: with X-ray contrast fluids we can examine protective and drainage function of ET. Contrast fluid is applied into nasopharynx and we

observe its retrograde movement into ET. Protective function of ET is normal, if contrast fluid cannot get into bone part of ET during swallowing. Drainage function can be examined by contrast fluid application into middle ear cavity and observation of its movement into nasopharynx. We can use scintigraphy or microendoscopy as well. Sonotubometry is usable in research, it is not accessible for clinical usage.

Otoscopy

Examination of eardrum by sight: we use ear mirror (speculum), microscope or otoscope. During otoscopy is essential to straighten the auditory canal, which has often sigmoid shape. We do so by pulling the auricle dorsally and externally in adults or cranially and externally in children. Patient´s position depends on its age and compliance.

Pneumootoscopy: use otoscope connected with ball, which can be used for pressure changes in auditory canal. If we tightly obdurate auditory canal we can observe mobility of eardrum with pressure changes.

Evaluation of otoscopic findings

We see Bezold´s trias on normal eardrum (prominentia mallearis, stria mallearis and light reflex). Prominentia mallearis is formed by processu brevis mallei, stria mallearis is formed by manubrium mallei, its end is tightly connected with eardrum and forms umbo membranae tympani. Light reflex comes up from light reflection, has triangle shape with apex in umbo and base in annulus. Eardrum can be divide by 2 imaginary axes (one goes through stria mallearis) into 4 quadrants: upper anterior, upper posterior, lower anterior, lower posterior.

- **Location of eardrum**: normal eardrum should be in neutral position with slightly protruding processus brevis mallei.

- 1. **Retraction of eardrum** is usually sign of underpressure or secretion in middle ear or both – mallear prominence expressively protrude and manubrium mallei looks like shortened and located more horizontally. If we evaluate eardrum retraction in pars tensa we use classification according to Sade. Pars flaccida retraction is classified according to Tose. Atrophic areas on eardrum can form so called retraction pockets. They are classified according to Charachon.

- 2. **Eardrum bulge** is noticeable mostly in its most supple parts – in epitympanum and upper posterior quadrant. Processus brevis mallei is indistinct. Eardrum bulge is caused by increased pressure in middle ear or by secretion or expansion.

- **Eardrum color:** is grey. Yellowish or bluish color can be caused by secretion in middle ear. Violet eardrum usually signs presence of blood in middle ear (hemotympanum).

- **Eardrum mobility:** is evaluated by pneumootoscopic examination or by tympanometry. Normal eardrum and ossicles are moving according to pressure changes in external auditory canal. Mobility is influenced mostly by secretion, negative pressure in middle ear or by fixation of ossicles. Increased mobility is found in atrophic eardrum or if ossicle string is

disrupted. Decreased mobility can be found in thickened eardrum, myringosclerosis, otosclerosis and in children during chronic secretoric otitis.

Subjective hearing examination

Conversation with patient

Valuable information from patient can be gained during history:

- Patient doesn´t understand even loud speech with reading the lips
- Patient understand loud speech with reading the lips
- Patient understand loud speech without reading the lips
- Patient understand silent speech without reading the lips

We can also notice pronunciation defects (in higher frequencies defects is sibilants pronunciation slurred), change of speech melody (more serious hearing defects) or head turning (asymmetric affliction). Especially in children can hearing loss cause uncertainty and fear, because they don´t understand what happens around them.

Classical hearing test

It is basic hearing test. It is made by loud or whispered voice (vox, vox sibilans – abbreviations V, Vs) Patient is turned by examined ear to doctor and by face to the assistant, who obstructs other auditory canal in whispered voice or din ear (for the deafeneding , insertion of olive or Barany´s device is necessary) in loud voice. According to distance we make a description, e.g.:

- 6m Vs 3m – patient hears whispered speech on right side from 6m and on the left side from 3m

Result is orientation and depends on patient compliance, doctor and assistant experiences and quality of given room (silence and sufficient length). Differences between loud and whispered test are now obsolete.

- Loud part is worse understood in case of lower frequencies affliction, because loud speech has majority of acoustic energy formed by vocals, which have significant formant structure with energy maximum between 100-1000 Hz.
- Whispered part is worse understood in case of higher frequencies affliction, because whispered speech has majority of acoustic energy formed by consonants, which have energy maximum between 2000-8000 Hz.

Sense of hearing test lies in examination of central hearing component. The more centrally is defect the more is hearing impaired and the less is worsened tone detection. Typical example is afasia, when tone audiometry is normal but understanding is severely afflicted. On the other way, understanding in light and moderate conductive hearing loss is relatively good.

Tuning fork examinations

Examination was made in the past by set of tuning forks of different frequencies. Now it is special test with restricted number of tuning forks (usually one). Their sense diminishes, but even now are good guide before tone audiometry and are priceless in understanding of diagnostics theory of conductive and perceptive hearing loss.

Rinne´s test: informs us, if hearing is better via air (fork placed in front of opening of auditory canal produce sound, which is transmitted throught auditory canal, eardrum, ossicles into inner ear) or via bone (fork placed on processus mastoideus vibrates with whole skull and os petrosum, where is located membranous labyrinth and where is stimulated inner ear). Energy needed for skull vibration and sense of tone is about 40 dB bigger than in healthy ear. Practical performance: vibrating fork is placed in front of auricle and patient tells us, when he stops hear it. Immediately after that fork is placed on processus mastoideus and patients tells us, if he hears it or not. If not, hearing system is intact or is present perceptive hearing loss. If yes, we reverse the course and if patient hears better through processus mastoideus, conductive hearing loss is present (but we cannot exclude by this mixed hearing loss).

Weber´s test: we place fork on top of head or on forehead. He tells us, in which ear he hears better.

- Conductive hearing loss – in afflicted ear
- Perceptive hearing loss – in healthy ear
- Mixed hearing loss – depends on both components

Result improvement can be reached by calibrated Weber´s test, when stimulus is lead to vibrator on the forehead from the audiometer. It allows examination of different frequencies and intensities.

Schwabach´s test: it is not usually done, because it compares hearing of patient and doctor

There are some specialized fork tests (Gelle, Cytovič, Frederici). However, they are suitable for only some diagnoses and are so complicated, that we don´t use them in children.

Tone audiometry

It is examination of clear tones hearing. It is made by audiologic nurse or doctor with help of audiometer, machine with generator of those tones. Sound is lead in air (headphone, speakers) or in bone (bone vibrator placed on processus mastoideus (see Rinne´s test). Examination is made in silent room or better in audiochamber (special, noise eliminated room). Patient responds to presented tone:

- Classic tone audiometry – pressing the button (from school children to adults)
- Classic tone audiometry in children – raising the hand (pre-school schildren)
- Tone audiometry with game – building tower from cubes, etc., usually with speakers (headphones are felt negatively in children)

- Behavioral audiometry – non-specific reactions as a blink, activity interruption, turning round on noise (age of 6-24 months)
- Audiometry with visual amplification – on base of conditional reaction, child is turning round on noise with supposed award (e.g. doll with blinking eyes), age of 6-24 months.

Result is graph – tone audiogram:

- Axis X means frequencies of tones in Hz (Hertz) – 125, 250, 500**,** (750), 1000, (1500), 2000, (3000), 4000, (6000), 8000 – in brackets are optional frequencies, most important frequencies are 500, 1000 and 2000 Hz.
- Axis Y means intensities of presented sound (in dB – decibel)
- Particular points determine threshold of hearing – tone sense on certain frequency induced by as lowest as possible intensity. It is marked by:
    - Air conduction – circle and cross, frequencies are connected with line
    - Bone conduction – brackets, frequencies are connected with dash line

There are other tests used for special audiologic test or for hearing-aid devices application – threshold for unpleasant listening, pain threshold, etc.)

Speech audiometry

Is group of examinations, where patient repeats words, whose are played in variable intensity. It is analogy of classical hearing test made in silent room or audiochamber and stimuli intensities are calibrated. Source can be microphone or record on e.g. CD. Typical is presentation of 10 words of same intensity. From number of correct answers is calculated percentage of intelligibility. Basic described values are 50% intelligibility – threshold of and the lowest value, where patient understand the most (usually 100%).

Possibilities of presentation are same as in tone audiometry:

- From speakers (so called free field)
- Into headphones
- Into bone vibrator (bilateral stimulation of inner ear)

Meaning of verbal audiometry:

- Evaluation on hearing-aid devices effect
- Hearing examination of pre-school children (words repeating or showing them on pictures is for children more simple than signalization of tone detection)
- Examination of speech intelligibility – in case of normal hearing according to tone audiometry, with help of special verbal groups

Objective audiometry

Tympanometry

Tympanometry is objective examination method evaluating changes of pressure in external auditory canal on the base of sound reflection from eardrum back to tympanometer.

Tympanometer is device, which emits sound waves towards eardrum, receives them and process reflected waves and pressure changes in external auditory canal. If pressures on both sides of eardrum are same, maximum of sound energy goes into inner ear (compliance (softness) of eardrum and ossicles is highest). The bigger is difference between both sides of eardrum, the more is compliance decreased and the more is admittance (rigidity) increased. The waves are reflecting back to the tympanometer so that tympanometer record several types of curves.

Tympanometric curves are usually evaluated by classification according to Jerger: basic curves are A, B, C.

- **Curve A**: is physiological with peak in zero pressure. Peak means value of actual pressure in middle ear (pressures on both sides of eardrum are same. Curve As (peak is lower then o.3ml on Y axis) is connected with increased rigidity of conductive system (e.g. otosclerosis). Curve Ad (has peak more than 1,2 ml above on Y axis) is connected with increased mobility of conductive system (eardrum atrophy, ossicle string disruption)

- **Curve C** means disorder of ET ventilation function. Peak on this curve is moved into negative pressure values (C1 from -100 to -200 daPa, C2 from -200 daPa)

- **Curve B** has no peak. It means, that eardrum reflects during different pressures same amount of sound. It is caused by increased rigidity of eardrum-middle ear system, usually caused by secretion in middle ear behind intact eardrum (OMCHS – otitis media chronica secretorica)

Positive admittance can be found in ambulance in acute otitis media, however, in case of this disease we don´t make tympanometry because of pain.

Majority of tympanometers can detect actual volume, where pressure change is occurring (external auditory canal, possibly middle ear cavity) and reveal small invisible eardrum perforation.

Modern tympanometers can examine even acoustic reflexes of middle ear muscles (m.stapedius – inervation from n.VII and m.tensor tympani – n.V). These muscles act as protectors of auditory apparatus against loud noise. If function of outer, middle and inner ear is intact, stapedial reflex can be noticed in intensities over 80 dB, reflex of m.tensor tympani in intensities over 100 dB. Reflexes are bilateral during unilateral stimulation. If reflexes are noticed in hearing loss, it means that disorder is located behind reflex arches. If stapedial reflex is noticed on levels up to 60 dB above threshold we consider it prove of recruitment. Decay reflex examine increased fatigue of hearing. If values of stapedial reflex decrease more than by 50% in 10 seconds, it means increased fatigue of hearing.

Otoacoustic emissions (OAE)

Sound produced by cochlea was described and measured by Kemp in 1978.

Otoacoustic emissions are generated as a nonlinear byproduct of cochlear biomechanic activity on the level of external cilliary cells. They are produced only preneurally and don´t show any ability to transmit sound. OAE examination is fast, non-invasive and objective.

There are 2 categories of OAE: spontaneous OAE (SOAE) or evoked OAE (EOAE). EOAE have important use in clinical praxis, they allow assessment of external ciliary cells function, which generates emission by mechanic touch as a response to sound presence. OAE are not noticed, if hearing loss is greater than 30 dB.

- Use: hearing screening in neonates, stimulation examination, examination in ototoxic therapy, perceptive hearing loss

Examination of evoked potentials

Examinations: ERA (electrical response audiometry) or AEP (auditory evoked potentials) are based on average EEG record. We evaluate electric potentials with different latency:

- Short – electrocochleography, BAEP (or BERA) up to 15 msec
- Middle – MERA, SSEP, VEMP – 15-100 msec
- Long – LAEP (or CERA), P300, MMN – more then 100 msec

For threshold determination we evaluate presence or latency of single waves. For auditory track description we describe latency, intervals and differences between sides.

- BAEP, **BERA** (brainstem AEP or ERA) evaluate potentials from brainstem. We describe 7 waves, the most constant is 5.wave, which originate from colliculus inferior, or waves 1 and 3.
- **Electrocochleography** evaluates potentials from cochlea – record is optimized maximally to the level of wave 1 of BAEP. The nearer to cochlea, the better the quality (best is transtympanic placement into round window)
- MERA (middle latency ERA) – classical examination of averaging is considerably afflicted by muscle artifacts (see VEMP), in general anaesthesia is practically unnoticeable. It is not used.
- SSEP (steady state evoked potentials) – hearing threshold is determined according to signal analysis (very fast and accurate examination, but we cannot describe auditory track).
- VEMP (vestibular evoked myogenic potentials) – is based on disadvantage of MERA. Muscle artifact is generated on the base of vestibule-spinal reflex during sacculus stimulation by great intensity (1000 dB nHL, 120 dB SPL). It is used in diagnostics of balance disorders.
- LAEP, **CERA** (long latency AEP, cortical ERA) evaluate potentials from auditory cortex (complex of waves P1N1P2). This examination is used especially in case of simulation, dissimulation, aggravation, because stimuli are same as in tone audiometry and allow objective examination till the end of auditory track. Waves P300 and MMN are used in

cognitive testing, which allow examination of processing auditory signal by auditory track and cortex (significant in examination of intelligibility.

Examination of vestibular apparatus

History – essential is description of circumstances, time, duration and character of problems development (balance disorders, nausea, vomitus, pressure in ear, tinnitus, hearing loss, collapse, headache).

Examination of vestibule-ocular reflexes

Examination of eye movements

Examination of spontaneous nystagm:

- Barthels´s glasses
- Frenzel´s glasses
- Videooculography
- Electronystagmography

Examination of semi-spontaneous nystagm:

- Position tests (slow position changes of body and head) – influence especially on utriculus and sacculus
- Positioning tests (fact position changes of body and head) – influence especially on semicircular canals
- Torsion test – diagnostics of vascularization defect of vertebral arteries
- Head shaking test

Examination of provoked nystagm:

- Temperature tests (with warm and cold water or air)
- Rotating and pendulum provocation

Examination of vestibule-spinal reflexes:

Orientation neurologic examination (everything with exclusion of visual fixation):

- Hautant – with arms raised forward and watching the deviation
- Romberg - standing (in variable stances) and watching the traction or fall
- Target pursue – point at nose with index finger
- Examination of adiadochokinesis (synchronic turning of hands)

- Walking on straight line
- Unterberg-Fucuda – 1 minute march on place, pathologic is more than 45 degrees deviation

VEMP – stimulation of sacculus with high intensity sound and registration of tonic responses of neck muscles

Posturography, stabilometry, craniocorpography – examinations of static or dynamic balance maintenance with help of objective methods

Imaging methods

Classical X-ray images are replaced by CT. However, Stenvers´s projection is suitable for patients with cochlear implant, because metal parts of implants can cause artifact during CT and make evaluation of CT difficult.

For detailed structure of temporal bone is most suitable HRCT (high resolution CT) in coronary and axial projection with scans 1-1,5 mm thin in bone algorithm.

For examination of inner auditory canal, brainstem and cerebellum area is suitable to make magnetic resonance – MRI. In comparison to CT MRI can reach better resolution of soft tissues and doesn´t irradiate patient. Disadvantages are: long examination time with necessity of anaesthesia in small children and non-cooperative persons and higher price.

Congenital defects of ear

Content

1. Apostasis auriculae
2. Microtia et atresia meati acustici externi
3. Fistula auris congenital
4. Genetic defects of hearing
    - 4.1 Structural anomalies of inner ear
    - 4.2 Autosomally recessive diseases
    - 4.3 Autosomally dominant disease
    - 4.4 Diseases bound to chromosome X
    - 4.5 Chromosomal aberrations
5. Congenital defects of auditory nerve
    - 5.1 Auditory neuropathy
    - 5.2 Aplasia or hypoplasia of auditory nerve

5.3 Demyelinization diseases

Apostasis auriculi

Congenital defect of anthelix is usually cause of auricle displacement.

Therapy: plastic correction. Ideal age for surgery is before school age (6 years)

Microtia et atresia meati acusticiexterni

Congential defect of auricle evolving (microtia) or missing auricle (anotia)is often combined with congenital defect of external auditory canal (stenosis, atresia). Incidence of other congenital defects is increased (defects of middle or inner ear) Auditory canal stenosis means, that it is narrower than 4 mm.

Diagnostics: CT, objective hearing examination (exclusion of congenital defect of middle and inner ear or auditory track)

Therapy: depends on examinations results and on hearing affliction extent (unilateral or bilateral affliction): hearin-aid devices, cochlear implant, tympanoplasty, plastic of external auditory canal or auricle

Fistula auris congenital

During auricle evolvement can develop in its surrounding fistulae and cysts. Most frequently we can find preauricular fistula with external opening placed between tragus and inner opening between cartilaginous and bone part of external auditory canal. Most frequent complication is inflammation, which causes secretion or sometimes swelling and erythema around fistula.

Therapy: ATB in case of inflammation, incision in case of abscess. Exstirpation of fistula in still state with sparing n.VII

Genetic defects of hearing

Genetic hearing defects can be congenital or gained, conductive or perceptive (SNHL – sensorineural hearing loss) or mixed, stabile, fluctuating or progressing, unilateral or bilateral, symmetric or asymmetric, syndromal (over 400 syndromes) or non-syndromal. They are characterized by audiology, age, progression and type of heritability (80% are autosomally recessive, 18% are autosomally dominant, 2% are bound to chromosomes)

Diagnostics: CT, objective audiometry

Structural anomalies of inner ear

20% of children with SNHL have CT anomalies of inner ear:

- **Michel´s type**: pyramid agenesis, outer and middle ear is usually normal. Can be confused with ossificating labyrintitis. Heritability can be autosomally dominant or recessive. Changes develop during 3.week of evolvement.
- **Mondini´s type**: develops in 6.week of evolvement. There are presented: 1 screw of cochlea, dilated ductus and saccus endolymphaticus, communication between scala tympani and vestibule. It is found in syndromes: Treacher-collins, Pendred, Waardenburg, Wilderwanck, Branchio-oto-renal, and in CMV infection. Affliction is unilateral or bilateral
- **Scheibe´s type**: wrong differentiation of Corti´s organ, we find malformation of tectorial membrane and Reisner´s membrane collapse. It is the most frequent type of affliction, found in syndromes: Jervell, Lange Nielsen, Usher, Waardenburg
- **Alexander´s type**: affliction of Corti´s organ in basal screw and dilated aquaeductus vestibule – typical bilateral affliction, cause perceptive hearing loss with fluctuating or progressing course, dizziness and balance disorders. Often is found in Pendred´s syndrome.
- **Malformations of semicircular canals**: most frequent are malformations of lateral canal

Autosomally recessive diseases

- Pendred´s syndrome: progressive SNHL, goiter as a result of iodine metabolism disorder
- **Jervell´s and Lange-Nielsen´s syndrome**: SNHL, lengthened QT interval on ECG
- **Non-syndromal hearing defects**: at least 20 loci were identified. Classification according to Konigsmark and Gorlin:
    - Congenital sever hearing loss
    - Middle hearing loss
    - Impairment with early onset

Autosomally dominant diseases

- **Waardenburg´s syndrome**: unilateral or bilateral SNHL (20% in type I, 50% in type II)
    - Type II (mutation of MITF gene): pigment anomalies (white spots, heterochroma iridis, vitiligo).
    - Type I (mutation of PAX3 gene): same as type I + dystopia canthorum
- **Stickler´s syndrome**: small jaws with palatoschisis (Pierre-Robin sy), myopia, cataract, hypermobility and enlargement of joints, arthritis in early adult, mixed or SNHL (80%), gene mutation COL2A1 on chromosome 12
- **Branchio-oto-renal syndrome (Melnick-Fraser):** ear appendages, neck fistulae, renal anomalies, hearing loss of different type

- **Treacher-collins syndrome:** Microtia and atresia of auditory canal, SNHL and CHL, hypoplasia of mandible, coloboma of lover eyelids, antimongoloid position of eyes
- **Non-syndromal hearing defec**ts: progressive hearing loss – variable age of onset, affliction of inner ear evolvement, disorders on many different frequencies

Diseases bound to chromosome X

- **Norrie´s syndrome:** SNHL, congenital or progressive blindness
- **Wilderwnack´s syndrome**: SNHL or mixed hearing loss + fusion of neck spondyls (Klippel-Feil sy), paresis of n.VI
- **Alport´s syndrome**: progressive SNHL, affliction of kidneys
- **Oto-palato-digital syndrome**: ossicular anomalies, hypertelorism, palatoschisis, small nose, fingers anomalies
- **Turner´s syndrome (X0):** SNHL or mixed hearing loss, gonadal dysgenesis, small stature, short neck. Multifactorial affliction.
- **Goldenhar´s syndrome:** preauriculary appendages, spine anomalies, epibulbar dermoid, coloboma of lower eyelid

Chromosomal diseases

Redundant or missing chromosomes can cause variable congenital defects.

- Down´s syndrome: trisomia of chromosome 21
- Patau´s syndrome: trisomia of chromosome 13
- Edward´s syndrome: trisomia of chromosome 18

Trisomia of another autosome is lethal.

Congenital defects of auditory nerve

Auditory neuropathy

Pathology in the area of synapse of inner ciliary cells and cells of auditory nerve

Diagnostics: objective hearing examination, MRI of brain

Therapy: cochlear or stem implantation in case of bilateral affliction. If it is not successful – sign language.

Aplasia or hypoplasia of auditory nerve

Diagnostics: objective hearing examination, MRI of brain

Therapy: in case of bilateral affliction stem implant with dubious effect, sign language

Demyelinization disease

See Neurology

Diseases of the external ear

Content

1. Cerumen
2. Erysipel
3. Eczema of external auditory canal
4. Otitis externa
    4.1 Otitis externa diffuza
    4.2 Otitis externa circumscripta
5. Perichondritis auriculae

Cerumen

Is produced by gl.ceruminosae of external auditory canal. Yellow-brown matter with fat particles can partially or totally obturate external auditory canal. It is getting dry and oxidates (getting hard and black). Can be cause of inflammation of external auditory canal or even hearing loss or tinnitus. Ceruman can be preferably removed by lukewarm water (cave thermal irritation of labyrinth). If we are suspicious of eardrum perforation we use boron water. Hard cerumen can be partially dissolved by oil preparations.

Erysipel

Streptococcal infectious disease of skin. It causes painful, circumscripted erythema.

Therapy: preferably PNC, wide spectrum ATB

Eczema of external auditory canal

Course: itching, increased sensitivity of auditory canal, discharge

Classification:

- Dry: increased production of skin epithelium
- Wet: blisters formation, discharge
- Mixed: combination of previous

Etiology:

- Contact: usually result of cosmetic preparations (soap, shampoo) usage
- Bacterial: in case of otitis externa, otitis media chronic mesotympanalis, otitis media chronic cum cholesteatomatae or otitis media acuta
- Atopic
- Seborrhoic

Therapy:

- Contact: change of cosmetics
- Bacterial: therapy of primary disease
- Atopic and seborrhoic: dermatological treatment

Otitis externa diffuza

Definition: inflammation of skin and submucous tissues of external auditory canal

Pathogenesis: usually result of bathing in contaminated or chlorinated water. It is disease of summer months

Course: pain, erythema and stricture of externa auditory canal. In severe cases: auditory canal impassability caused by soft tissues swelling, conductive hearing loss, increased body temperature, erythemaand swelling in periauricular area (dif.dg.acute mastoiditis: tympanometry, X-ray, CT).

Diagnostics: see Pathogenesis and Course. Pain increases by pressure on tragus or by auricle pulling.

Therapy: local ATB in unguent (chloramphenicol) or drops (Garasone, Otosporin), analgetics, in severe cases ATB generally

Complications: inflammation spread through Santorini´s fissures into skull base (otitis externa maligna) or into glandula parotis. Increased risk is present in patients with immunity defects or diabetes mellitus.

Otitis externa circumscripta

Definiton: affliction of small skin glands (gl.ceruminosae) or hair follicles of external auditory canal.

Course: painful erythema and swelling in cartilaginous part of external auditory canal (folliculitis, furuncle). Discharge after perforation or incision.

Diagnostics: see Course. Pain increases by pressure on tragus or by auricle pulling.

Therapy: incision, local ATB, analgetics, in severe cases ATB generally

Perichondritis auriculae

Cartilage inflammationdevelops usually after ear injury, or e.g. after insect bite. Without therapy cartilage suffers from insufficient nutrition from perichondrium which leads to cartilage destruction with permanent deformation of auricle

Therapy: ATB generally, punction, incision, drainage, remove destroyed cartilage.

Diseases of the middle ear

Content

1. Acute inflammations of middle ear
   - Otitis media acuta (OMA)
   - Otitis media acuta recidivans (OMR)
   - Myringitis acuta
2. Chronic inflammations of middle ear
   - Otitis media chronica mesotympanalis
   - Otitis media chronica secretorica
   - Otitis media chronica cum cholesteatomatae
   - Otitis media chronica adhesiva
3. Degenerative diseases of the middle ear
   - Otosclerosis
   - Tympanosclerosis
   - Myringosclerosis

Acute inflammations of the middle ear

Otitis media acuta (OMA)

Definition: mucosa inflammation of tympanal cavity and pneumatic system of temporal bone, followed by suddenly developed symptoms of acute infection.

Pathogenesis:

- Infection spreads most frequently from nasopharynx through Eustachian tube thanks to pressure gradient between middle ear cavity and nasopharynx (ET closed) or by snuffling (ET opened).
- Hematogenous infections during airways infection (flu, spála, neštovice)
- Infections of middle ear in case of tympanal perforation or another comunication between middle ear and outside environment

Etiology:

- Streptococcus pneumoniae (up to 60%)
- Haemophillus influenzae (up to 25%)
- Moraxella catarrhalis
- Staphylococcus aureus

After vaccination against H.influenzae and S.pneumoniae we suppose changes in percentage of specific pathogens.

Symptoms and diagnostics:

Initial stage:

- Subjective problems: airways infection, pricking or pain in the ear, worsened hearing, nausea, restlessness
- Objective findings:

otoscopy: accented veins, red tympanum, which is in its normal position – mallear prominence is distinct (processus brevis malei)

audiometry: normal or cinductive hearing loss

tympanometry: curve C. In neonates, toddlers and young children is necessary to differentiate congestion of the tympanum caused by other condition, e.g. by scream.

Stage of advanced otitis: in this stage appears bacterial superinfection on primary viral inflammation. Increased pressure of secretion leads to increased pain because of periferal sensitive nerves irritation. Risk of inflammation spreading to the bone with complications is also increased. If ET is opened, partial drainage of secretion to the nasopharynx is possible.

- Subjective problems: pain in the ear, hearing loss, nausea, vomiting (Arnoldś reflex in r.auricularis n.vagi is irritated). Small children are increasingly restless especially in horizontal position, beacuse of increased head and neck vascularization.
- Objective findings:

Otoscopy: bulge of the tympanum with maximum in dorsal upper quadrant (processus brevis malei cannot be differentiated)

Audiometry: převodní nedoslýchavost. In case of normal hearing Weber´s test contralaterally lateralizate into sick ear.

Tympanometry: isn´t made because of pain.

Stage of advanced otitis with perforation: increased pressure of secretion often causes tympanal perforation and drainage from middle ear cavity into external auditory canal. Infected secretion can cause inflammation of the external auditory canal.

- Subjective problems: hearing loss
- Objective findings:

Otoscopy: secretion in the auditory canal, maceration of the tympanum

Audiometry and Weber´s test: same as previous stage

Tympanometry: is impossible due to tympanal perforation

Therapy:

- Initial stage: therapy of airways infection, strip with boralcohol put on the tympanum, possibly ear drops. Analgetics, antipyretics, antihistaminics, fluids, vitamins, cold linen behind ear, increased position of the head. Examination after some time by general practitioner. If problems endure, examination by ENT specialist is needed.
- Stage of advanced otitis: paracentesis is fully indicated in dorsal lower quadrant. It lowers risk of complication and relieves from pain. After paracentesis pain diminishes and body temperature decreases to normal. Secretion from the ear stops in a week. Intensive local treatment is necessary after paracentesis. We reccomend repeated výplachy of external auditory canal with lukewarm bor water in accordance to intensity of secretion form middle ear, in first 3 days at least five times a day. Antibiotics are commonly not needed. During more serious course or another indicated cases amoxicilin is antibiotic of choice. We can give cefalosporins as well, in case of allergy macrolids. Control examination is made after 5 days after paracentesis. Patientes are told, that if problems are same or are even worse, they must visit specialist sooner. Hearing examination (in patients without compliance only tympanometry) is made after inflammation healing – commonly 14 days after first visit. Next control is planned according to results of audiometry or tympanometry - once per 1-2 months.
- Stage of advanced otitis with perforation: same as stage 2, but without need of paracentesis

Otitis media acuta recidivans (OMR)

Definition: rozvinutý acute middle ear inflammation (paraentesis, spontaneous tympanal perforation) three times in 6 months or four times in a year.

Course: possibility of otitis media chronica secretorica in coincidence (between recidives of acute inflammation of middle ear tympanometry B, eventually převodní nedoslýchavost), or without otitis media with secretion (tympanometry A and normal hearing)

Therapy: insertion of ventilation tube premeatuss other recidives of acute inflammation of the middle ear in most patients. In case we aren´t successful, imaging methods are needed (CT, X-ray) followed by surgical intervention (antromastoidectomy). Antibiotic premeatusion is ultimum refugium after imunological examination. OMR is one of the indications to vaccination against S.pneumonie.

Myringitis acuta

Definition: acute viral inflammation localized on tympanum itself

Pathogenesis: hematogenous infection during airways inflammation

Etiology: respiratory viruses

Course: pain of the ear during infection

Diagnostics: otoscopy – puchýřek on the tympanum

Therapy: paracentesis, therapy of airways infection

Chronic inflammations of middle ear

Otitis media chronica mesotympanalis

Definition: presence of eardrum perforation lasting at least 3 months with repeated secretion from middle ear cavity and conductive hearing loss.

Pathogenesis: malfunction of Eustachian tube, repeated middle ear injuries or eardrum injury. Influence of Eustachian tube malfunction type is unknown. In literature is written, that patients with open Eustachian tube have more often chronic mesotympanal otitis than patients with closed Eustachian tube.

Etiology: in bacteriological examination can we find E.coli, Proteus, Pseudomonas and others.

Course: repeated discharge from middle ear cavity into external auditory canal, often after water penetration into middle ear. In case of long lasting inflammation there is possibility of mucosal polyps and granulations development.

Diagnostics:

Otoscopy: eardrum perforation of variable range, commonly of kidney shape

Tympanometry: classical curves cannot be examinated, we can make only auxiliary function – Eustachian tube examination, volume measurement (helps differential diagnostics of retraction and perforation).

Hearing examination: commonly light or moderate conductive hearing loss

Therapy: local therapy for premeatusion of middle ear discharge, similarly as during acute inflammation of the middle ear, local ATB drops, or possibly ATB generally. Surgical therapy – removing of middle ear polyps and granulations. If patient is without middle ear discharge fro 3 months and the inflammation is cured, we call it a dry perforation of the eardrum (perforation membranae tympani or residua post otitidem) and as a definitive solution of this state is indicated myringoplasty. After closing the middle ear cavity by myringoplasty hearing is improved and life comfort of the patient is increased, because of premeatusion of repeated discharge from middle ear.

Otitis media chronic secretorica

Definition: secretion presence behind solid eardrum without signs of acute infection. If it last at least 3 months we assess it as a chronic inflammation. Presence of secretion up to 3 months indicate subacute secretoric otitis. Secretion can be purulent, mucous, serous or combinated.

Pathogenesis: malfunction of Eustachian tube or acute middle ear inflammations lead to structural and functional changes of middle ear and eardrum. In middle ear mucosa are formed glandular

structures, which are not present in normal healthy mucosa. Malfunction of ciliated epithelium leads to malfunction of ciliated transportation to the Eustachian tube. Result is accumulation of middle ear glands secretion or rest of secretion after acute otitis media. We can see atrophic spots and calcifications on the eardrum. Risk of retractions is increased in weakened parts of the eardrum.

Etiology: in middle ear secretion was found DNA of H.influenzae

Course: in the beginning is asymptomatic, later is present light or moderate conductive hearing loss. Possible is tinnitus and recurrent acute otitis media. Eardrum retraction in atrophic spots can lead to adhesions or cholesteatoma.

Diagnostics:

- Ototscopy: in the beginning can be normal otoscopic finding (grey, shiny and contoured eardrum. However, change of eardrum color by secretion is typical. Eardrum can be yellowish or bluish, in some cases we can see level of secretion. Reflex is fragmented, shortened or disappeared, eardrum is matt. Concavity of the eardrum lead to even more horizontal location of the stria mallearis. Retraction pockets are indication to surgical treatment.
- Tympanometry: curve B or C2
- Hearing examination: light or moderate conductive hearing loss

Therapy:

- Conservative: stimulation of palatal muscles (chewing gum), antihistaminics, local corticosteroids. Politzeration can be made only in patients without upper respiratory infect.
- Surgical
    1. Improving Eustachian tube function: sufficient Eustachian tube function can lead to improved mucociliated transport from middle ear cavity: surgical revision of nasopharynx and nasal cavity, improving of nose patency (by removing lymphatic tissue in nasopharynx in childhood. Lymphatic tissue can cause compression of nasopharyngeal entrance of Eustachian tube or can be the source of recurrent infections of nasopharynx). Especially in adult patients is necessary to exclude nasopharyngeal tumor.
    2. Improving hearing: by removing secretion from middle ear cavity and decreasing its production (myringotomy with secretion aspiration from middle ear or insertion of ventilation tube – risk of retraction pockets development is lower and aeration of middle ear cavity is ensured).

According to studies which assess effect of ventilation tube insertion - long term results of OMS therapy are not statistically different in patients with or without ventilation tube. However, benefits of ventilation tube are better hearing during disease and less extensive surgical interventions on temporal bone.

Otitis media chronica cum cholesteatomatae

Definition: presence of spinocellular epithelium in improper localization (in middle ear cavity in case of cholesteatoma)

Classification: according to development mechanism we divide cholesteatoms into congenital or gained. Accurding to localization we divide them into:

1. Tensa cholesteatoma
2. Flaccida cholesteatoma
3. Sinus cholesteatoma
4. Cholesteatoma with solid eardrum

Tensa cholesteatoma comes from upper frontal quadrant of eardrum, sinus cholesteatoma from upper dorsal quadrant, flaccida cholesteatoma from Schrapnell membrane. Cholesteatoma with solid eardrum can be congenital, posttraumatic or iatrogenous.

Pathogenesis:

- **Retraction theory**: long lasting ET malfunction and underpressure in middle ear lead to structural changes of middle ear mucosa and eardrum – glands formation, atrophy, calcification. Result is retraction pockets formation due to pulling atrophic areas medially. Sadé differentiated retraction pocket into 4 groups. Retractions in pars flaccid can be classified according to Tose. Cholesteatoma is formed, if epithelial cells from external layer of eardrum accumulate in those pockets. It happen often due to increased proliferation during chronic irritation. Retraction pocket forms matrix, which produces other epithelial cells and cause growth of whole formation. If fibrous membrane between epitympanum a mesotympanum is formed, it causes ventilation problems and underpressure in atticum and processus mastoideus, even if ET function is normal. This barrier isolating epitympanum is formed as a result of recurrent acute inflammations or repeated surgical interventions.
- **Proliferation theory**: In retraction pockets was proved deep papillary growth of flagstone epithelium of external eardrum layer.
- **Implantation theory**: applies especially during iatrogenous or posttraumatic cholesteatoma behind intact eardrum without retraction pocket. It supposes implantation of flagstone epithelium in tympanal cavity e.g. during paracentesis.
- **Metaplastic theory**: describe rare cases behind intact eardrum with adequate history. It supposes metaplasia of the middle ear epithelium to flagstone epithelium.
- **Congenital cholesteatoma**: besides in middle ear can be present e.g. intracranially. Congenital cholesteatoma can be named hamartoma.

Course: growing cholesteatoma causes destruction of surrounding tissues including bone with possible intratemporal and intracranial complications. Conductive hearing loss is frequent, but haven´t to be present even in case of total destruction of the middle ear bones, because cholesteatoma can well conduct sound. If inner ear is damaged by cholesteatoma (bone destruction, toxic damage), can be present vestibular symptoms or perception hearing loss. During infection can be present stinking secretion.

Diagnostics: otoscopy or microotoscopy, imaging methods (X-ray, CT), hearing examination

Therapy: surgical intervention in middle ear, small retraction cholesteatoms can be removed during microotoscopy. Cholesteatoma should be treated as soon as possible due to possible complications. We must count with possibility of recurrence.

Otitis media chronic adhesive

Definition: adhesions presence in middle ear as result of changes during chronic inflammation

Pathogenesis:

- adhesion of retracted eardrum with middle ear structures, usually in the area of long branch of incus, promontorium or epitympanum. Blood supply disruption of middle ear bones leads to their destruction.
- Fibrous strips formation, which causes bones fixation in middle ear as a result of chronic inflammation, presence of foreign body or postoperative.

Course: conductive hearing loss is present, often with signs of decreased mobility of bone string. Increase mobility is present, if bone string is broken or eardrum is atrophic. If risk of retraction is increased, risk of cholesteatoma development is also increased.

Diagnostics:

Otoscopy: retracted eardrum, retracted pockets or sometimes practically normal otoscopic picture

Audiometry: conductive hearing loss

Tympanometry: if bone fixation is present we find curve As or B, if breakage of bone string we find curve Ad.

Therapy: tympanotomy, tympanoplasty

Degenerative disease of the middle ear

Otosclerosis

Definition: degenerative affliction of temporal bone in white race (10%). Women are afflicted twice more than men. Disease frequency is lower in coincidence of morbilli vaccination.

Etiology: unknown, morbilli virus probably activates responsible gens (autosomally dominant heritability)

Pathogenesis: resorption of normal bone (osteolysis of osteocytes) and its replacement by sponge or sclerotic bone (unorganized bone with higher count of osteocytes and fibrous tissue). In 80-90% is pathology restricted on ventral edge of round window with calcification of ligamentum annulare. It can afflict cochlea and labyrinth as well. Pregnancy or hormonal changes can cause disease progression.

Symptoms: usually around 20.year of age:

- Slowly progressing, usually conductive hearing loss
- Paracusis Willisi: patient hear better in murmur
- Tinnitus or dizziness in more severe cases
- Syndrome “van der Hoeve” (otoscelrosis + osteogenesis imperfecta)

Diagnostics:

- History: heritability, pregnancy, hormonal changes and medication
- Otoscopy: usually normal eardrum. IN 10% Schwartze´s sign: black and blue color of eardrum caused by increased vascularization of promontorium
- Examination of hearing: tone audiometry: conductive hearing loss is in the beginning in lower frequencies, later even in higher. Carhart´s notch: decrease of bone conduction in frequency of 2000 Hz. In case of cochlear affliction is present mixed hearing loss in higher frequencies. During tympanometry we can find curve A or As.

Therapy: hearing-aid devices, surgery (stapedotomy, stapedectomy)

Tympanosclerosis

Definition: formation of thick fibrous tissue in middle ear usually with content of calcium salts

Etiology: unknown, middle ear inflammations, postoperative states

Symptoms: conductive hearing loss if middle ear bones are afflicted

Diagnostic: hearing examination, tympanometry (curve As or B)

Therapy: hearing-aid devices or tympanoplasty in case of conductive hearing loss

Myringosclerosis

Definition: formation of thick fibrous tissue in eardrum usually with content of calcium salts

Etiology: unknown, middle ear inflammations, after surgery intervention, paracentesis or ventilation tube insertion

Symptoms: conductive hearing loss if affliction is extesive

Diagnostic: hearing examination, tympanometry (curve As or B)

Therapy: hearing-aid devices or myringoplasty in case of conductive hearing loss

Inner ear diseases

Content

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2. Hearing loss caused by noise
3. Genetic hearing loss
4. Presbyacusis
5. Tinnitus
    5.1 objective tinnitus

    5.2 subjective tinnitus

Acute hearing loss

Definition: acute hearing loss with minimum of 30 dB on 3 neighbour frequencies in 3 days.

Pathogenesis:

1. viral – serous labyrintitis
2. impaired blood perfusion – increased risk in patients with hypercholesterolemia, hypertension, diabetes, age above 50 years, hypercoagulation factors
3. cochlear membranes damage – pyramid fractures, acoustic trauma, barotrauma, endolymphatic hydrops
4. immune affliction – autoimmune (with or without systemic autoimmune deisease, rheumatoid artritis, Cogan´s syndrome, Wegener´s granulomatosis, ulcerative colitis), or parainfection affliction of inner ear

Symptoms: hearing loss, speech understanding impairment, tinnitus, pressure in the ear, ear pain. In case of membranous labyrinth damage are present signs of balance disturbance – dizziness, nausea, sight impairment (problems with focusing on moving objects, or in fast change of vision)

Diagnostics, therapy, complications and consequences according to specific chapters.

Autoimmune diseases of inner ear

Definition: (fluctuating progressive) acute sensorineural hearing loss, with 15 dB decrease in 1 frequency or 10 dB decrease in 2 or more frequencies in several days. It reacts on antiinflammatory drugs, especially corticosteroids.

Pathogenesis:

- histamin induced vasodilatation on the base of I.type alergic reaction causes endolyphatic hydrops because of damaged ion and fluids exchange in endolymph (improving ear symptoms in alergics after antialergic drugs administration)
- settling of circulating imunocomplexes in inner ear arteries (e.g. in patients with systemic lupus erythematodes or Wegener´s granulomatosis)

- production of antibodies against antigens of inner ear (proved experimentally, but no test for clinical praxis exists)

Etiology: unknown (congenital, crossed antigenicity with wall of some bacteria)

Symptoms: usually bilateral hearing loss, tinnitus, pressure in the ear, rarely ear pain. In case of membranous labyrinth damage are present signs of balance disturbance – dizziness, nausea.

Diagnostics: audiometry, exclusion of retrocochlear hearing loss (BAEP, MRI), in case of balance problems is suitable vestibulologic examination, inflammatory markers (sedimentation, CRP), antinuclear antibodies, rheumatoid factor, circulating immunocomplexes, parts of complement, other serologic examination according to rheumatologist or immunologist consultation. Hearing improvement (15 dB on 1 frequency or 10 dB on 2 or more frequencies) after corticosteroid administration is essential for diagnostics.

Therapy: corticosteroids per os, intravenously or local aplication into middle ear (penetration to inner ear via round window). Not all patients respond to treatment. Not all of them, who respond, are without reccurence. Probability of success decreases with time from symptoms start. Efficiency borderline is about 30 days.

Complications and consequences: hearing loss or even hearing loss, balance problems. Long term corticosteroid treatment has strong side effects, which overweight possible profit from this therapy.

Labyrintitis

Definition: inflammatory disease of membraneous part of inner ear and/or labyrinth.

Pathogenesis:

- hematogenous dissemination
- passing from middle ear throught natural connections of middle ear and perilymphatic space (foramen ovale and rotundum), or throught perilymphatic fistula
- from central nervous systém (subarachnoideal space) throught internal auditory canal and aqueductus cochleae

Etiology:

- serous labyrintitis – bacterial toxins, mediators of inflammation, viruses (CMV, HSV1, mumps, rubeola, influenza, parainfluenzy, adonviruses, coxackie viruses, RS virus)
- syndrome Ramsay-Hunt (herpes zoster oticus) – reactivation of latent viral infection (varicella-zoster), often many years after primary infection

- Suppurative labyrintitis – most frequent agents of acute and chronic inflammations of middle ear (OMA), Neisseria meningitidis and others

Symptoms: dizziness, nausea, hearing loss, pressure in the ear, tinnitus, ear pain, febrils, meningeal signs, signs of respiratory infect, sight impairment

Diagnostics: otoscopy, hearing examination, meningeal signs, basic vestibulologic examination (nystagmus, Hautant´s test, Rombergś test, test Unterberg-Fucuda), eye examination, lumbal punction (serology, cultivation). In indicated cases CT, MRI or advanced vestibulologic examination.

Therapy: ATB in suppurative labyrintitis, antivirotics in indicated cases, corticosteroids, antiemetics, anitvertiginous drugs, hydratation. Surgical intervention in case of infection focus in the middle ear

Complication and consequencies: hearing loss, ossification of the cochlea (makes cochlear implantation imposible), infection spread to meninges, dysequilibrium (problem with balance even after considerable improvement of the dizziness)

See also Labyrintits in Otogenous inflammatory complications

Menier´s disease

Definition: hydrops (pressure change between endolymp and perilymph) of semicircular canals in inner ear without known etiology (after exclusion of all other causes in this chapter)

Classification:

Typical (presence of all symptoms based on affliction of cochlea and vestibular part of inner ear) and atypical

- hydrops cochlearis – only cochlea is afflicted (dizziness is not present)
- hydrops vestibularis – vestibular part of inner ear is afflicted (hearing loss is not present

Etiology: unknown, microvascular blocade is supposed, known are endocrine influence (more frequent onset in menses or menopausa)

Symptoms: fluctuating hearing loss, tinnitus, several minutes of rotatory vertigo, pressure in the ear. Symptoms are episodic.

Diagnostics: hearing examination, vestibular examination, electrocochleography and BERA. Glycerol or furosemide test (hydrops is confirmed if hearing improves after drug administration). MR of brain to exclude pathologic changes of acoustic nerve or brain. CT of pyramids if we have suspicion on perilymphatic fistula.

Therapy: diet (salt, caffeine and alcohol restriction), diuretics, corticosteroids (per os, intravenously or into middle ear), symptomatic therapy (antiemetics, sedatives). Destruction of inner ear (surgically or chemically – aminoglycosides injection into middle or inner ear) in case of repeated recurrences with bad quality of life.

Ototoxicity

Acoustic trauma

See Hearing loss caused by noise

Pyramid fracture

Hearing loss caused by noise

Clinically we differentiate 3 diagnostic units:

- acoustic trauma
- hearing loss cause by long lasting noise (especially professional) – influnce of recurrent and overpower noise in months and years
- socioacusis – hearing of city dwellers is worse than hearing of countrymen

Pathophysiology: temporary treshold shift (TTS) is caused by metabolic exhaustion of ciliary cells exposed to overpower noise. Lasting stress leads to microijuries – stereocilia affliction of inner and outer cilliary cells, their apoptosis, rupture of Reisner membrane, destruction of ciliary cells which all results in permanent hearing loss (PTS – permanent treshold shift). Affliction is biggest on frequencies of 3000-6000 Hz, often with greatest notch on 4000 Hz because of 2 reasons:

- protective function of reflexes of m.stapedius and m.tensor tympani in deep frequencies up to 1000 Hz
- interference of waves in basillary membrane on 4000 Hz according to Barány´s theory

Etiology: level of affliction and chance of improvement depends on noise dose (noise intesity x time of noise stress) Affliction is symetric in case of long lasting stress. In clashes (short sound of high intensity). Is affliction usually unilateral depending on direction of incoming noise. Hearing loss can develop after 8 hours in 85 dB noise. Double energy of sound (about 3 dB increase) cause similar damage in half of time. Persisten noise is worse than intermitent. Other factors are: individual endurance connected with genetic influence and global state of organism (higer risk is in diabetics, patients with hyperlipidemia, with cardiovascular diseases, with tobacco abuse or in patients who have ototoxic drugs).

Diagnostics: subjective audiometry, tympanometry with stapedial reflexes. Retocochlear hearing loss must be excluded (BAEP, MRI) only in case of asymetric hearing. Labortatory tests are useless.

Differential diagnostics: Simulation, dissimulation and aggravation must be excluded by cortical evoked potentials (LAEP, CERA) in case of professional diseases or money compensation.

Therapy: there is no proved effective therapy. Ve can offer corticosteroids, vasodilatating drugs or hyperbaroxytherapy with corresponding information about side effects.

Premeatusion: is the most important due to minimal or no effect of any therapy. Maximum levels of noise, which should be harmless (according to A filter):

16 h – 85 dBA

8 h – 90 dBA

6 h – 92 dBA

4 h -95 dBA

3 h – 97 dBA

2 h – 100 dBA

1,5 h – 102 dBA

1 h – 105 dBA

30 min – 110 dBA

15 min – 115 dBA

Genetic hearing loss

Presbyacusis

Changes of hearing connected with old age. We have 4 types according to Schuknech:

- sensoric – strophy of ciliary cells
- neural – atrophy of acoustic nerve and auditory pathway neurons (unlike others, it doesn´t afflicts hearing too much, but significantly impaires speech understandig)
- strial (metabolic) – atrophy of stria vascularis (maintain chemic and bioelectric balance in the inner ear)
- conductive (mechanic) – thick and rigid basillary membrane of the cochlea

Etiology: genetic predispositions, diabetes mellitus, arteriosclerosis, noise, ototoxic substances, stress

Diagnostics: hearing examination, other examinations only due to differential diagnostics

Therapy: there is no causal therapy. Premeatusion is important (elimination of etiological agents). Hearing-aid devices, cochlear implantates, FN systems and others, reading the lips.

Tinnitus

Definiton: perception of sound, whose source is no in patient´s surrounding, head or ears.

Objective tinnitus

We can hear or register by any other way, or logically connect with physical source of sound.

Etiology: the most frequent cause is vascular (turbulent flow in vertebral arteries and so called "jugulary venous noise"), less frequent is muscular (palatal myoclonus nad degenerative neuromuscular diseases – see neurology)

Diagnostics: examination of auditory canal and middle ear, hearing examination, examination of stapedial reflexes, Eustachian tube tests. If we suspect vascular etiology, angiography can be made.

Therapy: pathology of auditory canal and middle ear removing, tenotomy of m.tensor tympani in case of palatal myoclonus, embolization in case of vascular etiology.

Subjective tinnitus

Source of sound doesn´t exist – it is a phantom and patient is aware of nonexistence of the source (appropriatly to its intelect, state and age). It ussually is simple sound (e.g. whistle).

Etiology: it follows majority of disease in this chapter. On any situation we don´t sense any sound, compensatory sound perception develops – tinnitus. Tinnitus can develop as well in case of auditory nerve or auditory pathways damage due to signal desynchronization .

Diagnostics: ENT examination (objective tinnitus exclusion), hearing examination, comparison and tinitus masking in sounds from audiometer

- Murmur is typical for disease of auditory canal, middle ear and cochlea (in the apex).
- Whistle is typical for basocochlear diseases
- Fizzle is typical for diseases of external auditory canal, middle ear, auditory nerve and auditory pathways
- Tinnitus caused by inner ear diseases can be better masked than from auditory nerve aor auditory pathways

Objective diagnostics of subjective tinnitus is in expermient made by functional imaging methods – PET or fMRI

Therapy: causal therapy accurding to etiological agent, „tinnitus retraining therapy" (abreaction, autosuggestion, masking by šumy, etc.)

Differential diagnotics: halucinations are differentiated according to aspect of patient on sound source and by character of the sound (as a running locomotive)

Tumors of the ear in children

They make only very small % of children tumors of neck and head (1,5% of primary tumors). Embryonal development can explain some patological states. Os petrosum ossificates enchondrally and is completely formed at birth. Squama, mastoids and os tympani ossificate endesmally and develop partially after birth. Theri active growth centers are predispositing factors for mesenchymal tumor development.

Content

Congenital malformations and paraneoplastic processes

Histiocytosis X

Ear symptoms: otorhea and retroauricular swelling, rarely ear pain. In otoscopy can be seen polyp, granulation tissue, eczema of auricle and auditory canal. Conductive hearing loss is present in case of auditory canal and middle ear infiltration by soft tissue. Perforation of eardrum in secondary infection. Perceptive hearing impariment and vertigo are rare and are present in labyrinth destruction and n.VII paresis.

Premeatusion of recurrence: control otoscopy once in a month, control CT once in a year

See also: Histiocytosis X in Tumors an expansive processes in children

Choristoma

Most frequent is salivary choristoma of the middle ear. It is related to abnormalities of n.VII or ossicles in patients vith congenital anomalies of external ear and face. In children under 5 years is afflicted usually left ear (but can be afflicted both). It grows slowly and cause conductive hearing loss.

See also: Choristoma in Tumors an expansive processes in children

Hamartom

The most frequent is slowly growing extracanalicular osteoma of mastoids and squama. Symptom is slowly worsen conductive hearing loss

See also: Hamartoma in Tumors an expansive processes in children

Teratom

The most frequent is auro-nasopharyngeal hairy polyp.

See also: Teratoma in Tumors an expansive processes in children

Cystic lesions of petrous bone apex

Are probably results of obstruction of petrous bone pneumatizations in children.

- congenital epidermoid (usually found in adolescents)
- cholesterol cyst (granulomatous lesion containing cholesterol crystals, bleeding is essential factor for developmet)

Symptoms: conductive hearing loss caused by OMS because of ET obstruction, headache because of bone destruction and meningeal irritation, diplopia from n.III and n.VI damage, hyperestesia in face (n.V), dizziness if labyrinth veins are damaged

Diagnostics: CT, MRI

Differential diagnostics: cholesteatoma

Therapy: surgical, marsupialization of cholesterol cyst and exstirpation of epidermoid cyst.

Children myofibromatosis

Temporal bone is usually one of many afflicted bones

See also: Children myofibromatosis in Tumors an expansive processes in children

Fibrous dysplasia

Temporal bone affliction: external auditory canal stenosis, retroauricular swelling, displacement of auricle, development defects of ossicles or inner ear

See also: Fibrous dysplasia in Tumors an expansive processes in children

Benign tumors

Neurinom n.VIII

Definition: slowly growing, usually unilateral, intracranial tumor from Schwann cells afflicting vestibular (95%) or cochlear (5%) part of nerve. 80% of tumors is located in pontocerebellar angle.

Incidence: 0,7-1,0/100 000

Etiology: coincidence with neurofibromatosis type I (unilateral) and II (bilateral)

Symptoms:

- most frequent is asymetric sensorineural hearing loss from cochlear nerve damage (slowly developing) or vascularization damage (acute onset)
- tinnitus, dizziness, balance disturbances, headache
- malfunciton of n.VII
- can be asymptomatic (even in big tumors)

Diagnostics: MRI with gadolinium contrast, hearing examination (subjective and objective methods)

Therapy: according to age, size and symptoms:

- follow up
- Leksel gamma knife
- Microsurgery

Adenoma

It is present especially in adults, in children is rare. Adenoms are nonvascular, gummy fromations, which can fill up whole middle ear and can spread to mastoids. They often tightly adhere to the mucosa, eardrum and ossicles. Bone destruciton is a sign of malignancy.

Symptoms: progressive unilateral hearing loss with sense of fullness in the ear and with tinnitus

Therapy: complete resection. Recurrence is rare.

Paragangliom

Synonym: chemodectom, glomus tympanicum (jugulare) tumor

Definition: red vascularizated sperical tumor developing from neuroectoderm.

Pathogenesis: develops from paraganglions beside vena jugularis and near Jacobson´s and Arnold´s nerve. More often is in women in 5.decade, but some cases are known even in small children. There is hereditary form as well.

Classification:

- glomus tympanicum tumor develops in middle ear from plexus tympanicus (Jasobson´s nerve)
- glomus jugulare tumor develops in the area of jugular bulbus

Symptoms:

- unilateral pulse tinnitus, sense of fullness in the ear
- conductive hearing loss, immobility of ossicles
- large tumor can cause bleeding, pain, perceptive hearing loss, dizziness and lesions of head nerves (n.VII)
- glomus tympanicum usually doesn´t cause bone destruction
- glomus jugulare is often connected with demineralization or bone erosion
- sweating, hyperactivity, restlessness, tachycardia, palpitations, hypertension can be caused by katecholamine production (diagnostics – vanil-almond acid in the urine)

Diagnostics:

- otoscopy: red and violet tissue behind the eradrum. Brown´s sign – overpressure during pneumootoscopy whiten eardrum and stop pulsation, if tumor is in contact with eardrum
- tympanometry: higher impedance and higher pulse of synchronic wave
- audiometry: conductive, rarely perceptive hearing loss
- imagign methods: CT, MRI, arteriography (embolization)
- laboratory tests: vanil-almond acid in urine

Differinetial diagnostics: aberrant a.carotis interna, aneurysm, high bulbus jugularis, perfiftent stapedial artery, hemoragic secretion

Therapy:

- surgical (laser), alfa and beta blockers if katecholamins are produced. Embolization is possible.
- Radiothreapy

Prognosis is bad.

S*ee also: Paragangliom in Tumor in the area of external neck*

Malignant tumors

Rhabdmyosarcoma (RMS)

It is the most frequent malignant ear tumor and temporal bone in children. In middle ear is most frequent embryonal RMS.

Symptoms: serohemoragic discharge from ear, non-suppurative granulations, polyp or tumor in exteranl auditory canal, neurologic symptoms (n.VII). If petrous bone is afflicted, ear patology is often minimal. Headache, paresis of n.VI, trigeminal neuralgia and Horner´s syndrome are still present. RMS is highly aggresive and cause local destruction. He has distant hematogenous metastases in 20% in the time of diagnostics (lungs, bones, bone marrow), regional lymphatic nodes are afflicted less.

Diagnostics: ENT, neurology, CT, MRI, lumbal punction, biopsy, X-ray of chest and bones, liver and spleen examination, bone marrow punction

Therapy: depends on localization and state, combined surgery, radiotherapy and chemotherapy

Prognosis: average survival is 7-12 months in case of ear RMS

See also: Rhbdomyosarcoma in Tumors and expansive processes in children

Other sarcoms: fibrosarcoma, malignant fibrous histiocytoma, iposarcoma, mesenchymal chondrosarcoma and extraskeletal Ewing´s sarcoma are rare

Leukemia

Acute lymphatic or lymphoblastic leukemia afflicts ear and temporal bone in 20% of patients. Leukemic infiltrates can cause hemoragic ulcers in middle ear and in external auditory canal, thickening of eardrum and mucosa, damage of n.VII and n.VIII, hearing loss and dizziness.

Lymphoma

Primary is rare. Non-Hodgkin lymphom can afflict middle ear and mastoids.

Other malignant tumors

Spinocellular carcinoma, melanoma, tumors of salivary glands (adenocarcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma) are extremely rare in chidren

Ear injury

Content

Auricle injury

Are divided into open and closed (blunt). Open injuries must be revised, desinfected and foreign bodies must bez removed. Skin suture is made in local or general anaesthesia according to injury extension and patient´sstate. ATB are given generally.

Othematoma

In case of blunt injury (combat sports) hematoma is formed usually on external side of eardrum, where is perichondrium coneccted to auricle less tightly than on posterior side. Inflammation of auricle cartilage can develop in case of open wound or infected hematoma (perichondritis auriculae). Without therapy is cartilage destroyed and auricle deformed because of cartilage nutrition defect.

Therapy: generally ATB, punction, incision, drainage, destroyed cartilage removal

Auditory canal injury

It happen most frequently during personal hygiene, when manipulating with foreign body.

Symptoms: pain, bleeding. Diagnostics: otoscopy

Therapy: local ATB

Injury of eardrum and middle ear

It happen most frequently during personal hygiene, when manipulating with foreign body, in pyramid fractures and in case of barotrauma (sudden air pressure change in the auditory canal).

Symptoms: pain, bleeding, hearing loss, tinnitus

Diagnostics: otoscopy, CT

Therapy: ATB, myringoplasty, tympanoplasty

Premeatusion: don´t inserc foreign bodies into auditory canal

Inner ear injury

See Pyramid fracture and Acoustic trauma

Pyramid fracture

It is usually present in polytrauma (car accidents, falls)

Diagnostics: ENT examination, otoscopy, CT of pyramids, hearing examination, neurological examination, examination of face and auditory nerve funciton. Liquorrhea can be proved by presence of beta-2-transferrine

- longitudinal fractures of pyramids: during otoscopy we usually find eardrum perforation and liquorrhea from it

Therapy: ATB i.v. (with penetration to the liquor), let the liquor flows out of ear, don´t obstruct auditory canal (risk of infection). After state of patient gets better, we can reconstruct the eardrum and middle ear (tympanoplasty)

- transverse fractures of pyramids: eardrum is usually intact. Inner ear affliction cause perceptive hearing loss or possibly vestibular symptoms. Affliction of midlle ear can be diagnosed during otoscopy – violet eardrum (hemotympanum) signs presence of blood with possible tinge of liquor in the middle ear. Liquor can flow out via ET into nasopharynx (paradox otoliquorrhea)

Therapy: ATB, after state of patient gets better, we can reconstruct middle ear (tympanoplasty)

- Oblique fractures of pyramids: combination of the previous two types

Otogenous inflammatory complications

Content

Intratemporal complications

Are most frequent from all complications of middle ear inflammations. If bone is destroyed, abscesses can develop (Besold´s, Mourne´s, Mouret´s,…) and inflammation can spread into skull base and to the surrounding tissues and into intracranium.

Acute mastoiditis

Definition: mucosa and bone inflammation of pneumatic systém of temporal bone

Symptoms:

- erythema and swelling in retro(peri)auricular area
- displaced auricle
- descent of posterior upper wall of auditory canal
- conductive hearing loss
- signs of middle ear inflammation

Diagnostics: Decontoured eardrum or discharge out of middle ear can be found during otoscopy. Pneumatic system of temporal bone is destroyed on CT or X-ray.

Therapy: antromastoidectomy, ATB

Chronic mastoiditis

Usually follows chronic or recurrent middle ear inflammations. It causes increased temperature, chronic ear pain and sensitivity above mastoid processus.

Therapy: antromasoitdectomy, ATB

Paresis of n.VII

Is usually result of toxic damage on face nerve in often dehiscent Fallopi canal.

Diagnostics: see Differential diagnostics of n.VII paresis.

Therapy: antromastoidectomy, ATB, corticosteroids, nootropics, rehabilitation

Petrositis

Is infalmmatory affliction of pneumatized systém of petrous bone. It causes Gradening´s syndrome (n.VII paresis, trigeminal neuralgia, discharge out of middle ear)

Therapy: antromastoidectomy, ATB, corticosteroids, nootropics, rehabilitation

Labyrintitis

Can be caused by toxic products of inflammation (fistula of lateral semicircula canal in cholesteatoma, toxin transfer thorugh round window in acute inflammation) – serous or toxic labyrintitis. Or infectious agent can penetrate into inner ear and cause diffuse inflammation – suppurative labyrintitis. Serous and suppurative labyrintitis can both be diffuse or circumscripted, acute or chronic. End stage of chronic labyrintitis is sclerosis of labyrinth. We find sensorineural hearing loss, dizziness, balalnce loss, nystagmus, nausea, vomiting.

Diagnostics: hearing examination including tuning fork tests (Weber´s test) and vestibular apparatus, imaging methods of temporal bone

Therapy: antromastoidectomy, ATB, corticosteroids

See also: Labyrintitis in Disease of the inner ear

Intracranial complications

- meningitis – antromastoidectomy is indicated
- abscesses – epidural, subdural, brain, cerebellar – antromastoidectomy or neurosurgical intervention with abscess evacuation is indicated
- tromboflebitis – sigmoideal sinus is usually afflicted. Trombosis afflict emissariium mastoideum as well and this causes increased palpable sensitivity during pressure on mastoid processus (Griesinger´s sign)

Therapy: antromastoidectomy, hematological therapy (trombolysis) if surgical intervention fails – trombus evacuation

- hydrocephalus – obstructive in chronic tromboflebitis

Differential diagnostics of ear diseases

Content

3.1 Perpheral vestibular syndrome
3.2 Central vestibular syndrome
4. Differential diagnostics of n.VII paresis

Differential diagnostics of hearing problems

Classification:

- conductive – see diseases of outer and middle ear
- sensorineural (perceptive) – cochlear nad retrocochlear – see diseases of inner ear and auditory nerve
- mixed – conductive + perceptive
- central – see neurology

Differential diagnostics of ear pain

Definiton: ear pain is subjective feeling determined by patient and localized around ear. In children non-speaking patients we presume ear pain on the base of local and global symptoms and clinical finding. The most frequent cause is inflammatory affliction of outer and middle ear. Children have ear pain usually in acute otitis media. They form about half of patients in ENT ambulances.

Classification:

- otogenous: often sharp, localized into external auditory canal or into its very proximity
- non-otogenous: usually blunt, localized in whole area of ear and in addition the pain is present in another part of head or neck. This type of pain is mediated by neck nerves.

Otogenous pain

- Infection:
    - Auricle inflammation (erysipel, perichondritis)
    - Auditory canal inflammatioon
    - Obturating cerumen
    - OMA
    - Chronic otitis media
    - Complication of middle ear inflammations
    - Herpes zoster oticus
- Injury
    - Auricle injury
    - Foreign bodies
    - Barotrauma, acoustic injury
    - Burns and frostbites of auricle
    - Fractures of temporal bone
- Tumors
- Hydrops of the inner ear (M.Meniere)

Non-otogenous pain

Pain transposed from swallowing or respiratory pathways and soft tissues of neck:

- Odontogenous (tooth decay, stomatitis, teeth growth,…)
- Injury of facial skeleton (fractures of mandible joints, mandible luxation)
- Inflammations of oral cavity and pharynx
- Foreign body in pharynx
- Status post tonsillectomiam et adenotomiam
- Tumors of pharynx and larynx
- Neck lymphadenitis
- Parotitis
- Injury of neck spine
- Processus styloideus elongatus
- Rhinosinusitis
- Other (psychogenic, megrim, skull base tumor, neuralgia of n.occipitalis major, n.VII, ggl. Pterygopalatinum

Differential diagnostics of dizziness

Peripheral vestibular syndrome

Peripheral vestibular apparatus affliction (semicircular canals, acculus, utriculus) and vestibular part of auditory nerve

Etiology:

- labyrintitis
- hydrops (M. Meniére)
- neuronitis of n.VIII
- kinetosis
- benign position paroxysmal vertigo (BPPV)
- paroxysmal peripheral vertigo – mostly in patients with perceptive hearing loss, or changes in blood perfusion of vertebral arteries (degenerative disease of spine)

Symptoms: mostly acute onset of rotatory vertigo, balance loss connected eith nausea and vomiting. In unilateral affliction are present typical directional deviations. It lasts usually minutes (exceptionally days), fast relief from symptoms, good central compensation.

Diagnostics: vestibular examination, hearing examination, neurological examination, MRI of brain, CT of pyramids

Therapy: causal according to etiology. Symptomatic (sedatives, antihistaminics, antiemetics, mineral balance)

Central vestibular syndrome

Affliction of brain stem, cerebellum and other brain structures participating on balance maintenance.

Etiology: CNS affliction – see Neurology

Symptoms: dysequilibrium (balance uncertainity) rather than dizziness, slow onset, without nausea and vomiting. Lasts days to years. Balance deviations are nonspecific. Relief is very slow or no at all. Central compensation is not possible.

Therapy: causal see Neurology, Neurosurgery. Rehabilitation is eesential in residual affliction.

Differential diagnostics of n.VII paresis

We must exclude cause of n.VII paresis developed as a result of pathology in ENT area.

Classification:

- central
- peripheral

Most frequent causes of peripheral paresis:

- Bell´s palsy – idiopathic, commonly result of viral infection
- Ear pathology – inflammations (OMA, mastoiditis, cholesteatoma), fractures of pyramids, tumors
- Parotis pathology – malignant tumors, injury, iatrogenous affliction (surgery)

Diagnostics:

- examination of n.petrosus major function – Schirmer´s test (made by ophthalmologist)
- examination of n.stapedius function – tympanometry (stapedial reflex)
- examination of chorda tympani function – taste in first two thirds of tongue

Therapy: according to primary disease, ATB, corticosteroids, nootropics, rehabilitation. Antromastoidectomy is indicated if inflammatory affliction of middle ear is present.

Basics of ear surgery

Content

1. surgical approaches into temporal bone
2. Sanitation surgery
    2.1 Antromastoidectomy
    2.2 Atticotomy
    2.3 Atticoantromastoidectomy
3. Reconstruction surgery – tympanoplasty

myringoplasty

tympanotomy

Surgical approaches into temporal bone

- retroauricular – skin incision about 5mm behind auricle
- endaural – skin incision in incisura heliotragica
- endomeatal – via meatus acusticus externus
- combined

Sanitation surgery

Antromastoidectomy

Pneumatization of processus mastoideum removal with communication enlargement to eardrum cavity (aditus ad antrum mastoideum) from retroauricular incision. Most frequent indication is mastoiditid and other inflammatory complications. In young children, who doesn´t have developed pneumatization, it is called antrotomy.

Atticotomy

Atticum surgery (epitympanum) from endaural approach. Most frequent indication is lesser cholesteatoma.

Atticoantromastoidectomy

Is indicated in case of extensive cholesteatoma from retroauricular or preaural approach. In contains sanation of atticum and pneumatization of processus mastoideus. Dorsal upper wall of bone part of external auditory canal has to be usually removed as well. If auditory canal is reconstructed or dorsal upper wall of auditory canal wasn´t removed we call it a closed technique (it usually demands sekond-look operation up to 1 year). In other cases we call it a open technique. Classification according to Wullstein is now obsolete. For ossicles string reconstruction are used artificial materials (metal, plastic) – piston, PORP, TORP.

Myringoplasty

Reconstruction in case of eardrum perforation. It is the simplest tympanoplasty. Small perforations can be resolved by removing margine of perforation and cover (silk, cigarette paper). For reconstruction of larger perforation we use autolognous materials. Now is prefered „hard“ chondroperichondral graft from tragus or dorsal part of auricle. Transplant stays in position of former eardrum (incisura tympanica).

Tympanotomy

Reconstruction of ossicles can be made after skin incision approx. 2 mm before eardrum and lifting off the eardrum. Autolognous material have disadvantage in form of insufficient nutrition and that´s why they can be destructed. Artificial materials are better (they do not destruct).

Comments to nose embryology

Nose placods are formed in third week on frontal prominence (olfaction area), which deepen and form olfaction sockets and olfaction vesicles.

- Frontal prominence is divided by 3 notches into 2 medial and 2 lateral prominences. Medial prominences deepen and turn caudally, so they separate olfaction socket from primitive oral cavity – membrana bucconasalis.
- Cranial prominences (upper jaw) grow together with lateral prominences of nose (line is base of sulcus nasolacrimalis) and with medial prominences (entrance to olfaction socket is formed and primitive nose entrance is circumscribed)
- After that both medial prominences grow together and they are base of philtrum. Lateral prominences form lateral parts of nose and medial parts of face.
- Dorsum nasi develops from unpaired area triangularis located above medial nose prominences. This way formed flat middle part of face rises with subsequent growth. Simultaneously grow together prominences of first branchial arch for upper and lower jaw and by this eyes are moved medially.
- Buconasal membrane rips in 4. Week, so that primitive choans are formed. All olfaction sockets cause in advance prominences in form of sulcus vomeronasalis jacobsoni, which later closes itself into canal inserted into nose septum.

Clinical anatomy and physiology of the nose

Nose is divided into external nose and nasal cavity. Nose is connected with system of paranasal sinuses functionally and anatomically.

Content

1. External nose
2. Nasal cavity
3. Paranasal sinuses
4. Function of the nose and sinuses

External nose

Has shape of three-sided pyramid. It is formed by bone and cartilaginous skeleton.

- Bone skeleton is formed by processus frontales maxillae and ossa nasalia
- Cartilaginous skeleton is formed by allae nasi, crus mediale cartilaginis alaris, cartilage alaris major, cartilage nasi lateralis

Skin of nose is typical of high number of sebaceous glands. In bone part is mobile, while in cartilaginous part is strongly connected to cartilages. Nose muscles work as a sphincters and opener of nostrils.

- Muscles inervation: n.facialis
- Vascularization: external part of nose is perfused by a.facialis, blood is drained away by v.facialis and v.angularis (without valves), which leads to inner canthus and is connected with intracranial vascular system. During inflammations of external nose and upper lip (trigonum mortis) there is risk of infection spread to the cavernous sinus.

Nasal cavity

It is divided by septum into two single cavities. It is covered by mucos with respiratory epithelium. Mucosa of nasal cavities is very resistant and adaptable against bacterial or viral infection as well as against physical and chemical agents. Secretion glands and calyciformm cells form two-layer film: inner (serous), where cilia oscillate and outer (mucous), where dust and dirt are captured. Dust from frontal part of nasal cavity is moved by ciliary oscillation into choanes in 20 minutes .

Sensitive inervation in nasal cavity is maintained by I. and II. branch of n.V.

Vascularization is from a.ethmoidalis anterior and posterior and from a.sphenopalatina

- Medial wall is formed by nasal septum, which has cartilaginous character in frontal part (cartilage alaris major, cartilage septi nasi) and bone character in posterior part /lamina perpendicularis ossis ethmoidalis, vomer)
- Lower wall is formed by hard (processus palatines maxillae, lamina horizontalis ossis palatinae) and soft palate
- Lateral wall: there are 3 nasal conchae protruding from it – lower, middle and upper concha. They divide nasal cavity into 4 nasal meatuss – upper, middle, lower and common. Ductus ethmoidalis leads to lower meatus. Duct from maxillary and frontal sinuses and from frontal ethmoides lead to middle meatus. Sphenoidal sinus and posterior ethmoids lead to upper meatus. It is formed by processus frontalis maxillae, facies nasalis maxillae, os lacrimale, os ethmoidale, lamina perpendicualris ossis palatinae, processus pterygoidei ossis sphenoidalis
- Upper wall is formed by sphenoidal bone (corpus ossis sphenoidalis) and skull base – lamina cribriformis ossis ethmoidalis. Through it goes filla olfactoria from region olfactoria into intracranium.

Paranasal sinuses

- Frontal group: maxillary, frontal, and anterior ethmoids lead to middle nasal meatus
- Posterior group: posterior ethmoids and sphenoids lead to upper nasal meatus

Mucosa of sinuse is formed by cilliary epithelium. Řasinky oscillate towards cavity entrance and maintain cleaning of the cavity. Size and extent of pneumatization is very individual. Development of paranasal sinuses starts in fetal stage. Newborn has ethmoids, subsequently with child growth are forming other sinuses (maxillary to the 2.year, frontal to the 6.year, sphenoidal around 10.year). Definitive shape and size have in adult age.

Function of the nose and paranasal sinuses

- **Immunologic**: mucosa contains lysozyme, secretion immunoglobulins, heparine cells. Nasal mucosa removes noxious agents by displacement, dilution, neutralization and isolation. Cleaning starts in nasal entrance, where bigger particles are captured on nasal hairs (vibrissae) and smaller particles are adsorbed on mucosal layer and by ciliary motion are moved towards nasopharynx. Transfer from nasal entrance to choans last physiologically about 20 minutes. Secretion from glands and calyciformis cells dilute captured noxious agents and neutralize by enzymes as well. Plasmatic cells producing antibodies are contained

in lamina propria mucosae. They can by incorporated by macrophages together with antigen. Other cells are histyocytes producing vasoactive substances. Immunoglobulins play important role for nasal mucosal defence, especially the IgA secretion ones.

- **Regulating**: warm up, moisten and removes filth from inhaled air. 70% of inhaled air goes through lower nasal meatus. Inhaled air is in nasal cavity warmed up, saturated by vapor and solid particles are captured. Warming up is done by countercurrent system (vascularization of nasal mucosa and cavernous system). Moistening is provided by plentiful secretion of serous glands and by vaporization from mucin and tears. Saturation (by vapor) is about 80%.
- **Protective:** sneezing is reflex from nV irritation by endogenous vasoactive substances or by external chemical or physical stimulation. Nasopulmonary reflex is caused by irritation of olfactory and trigeminal nerves and it effector is n.X and respiration muscles inervation. Respond is cough or glottis closure. Nose blowing is non-physiological process, which replaces natural self-cleaning ability of nasal mucosa in case of pathological affliction. Respiratory epithelium of airways is impervious for common bacteria, if natural defense mechanisms are functioning. Previous mucosa damage is necessary for bacterial infection, usually by viruses.
- **Articulatory:** as a part of resonant space participate on man´s physiognomy
- **Olfactory and reflective:** peripheral olfactory analyzer is located in anterior upper part of nasal cavity vault – regio olfactoria. It receives stimulations in form of smells and stenches – it has protective value in differentiating the noxious agents. Man is able to differentiate more than 10 000 smells and stenches.

Breathing through nose goes physiologically against resistance, which is important in childhood for forming of oral cavity, muscle and pectoral skeleton development. That´s why is so important right nose breathing in children.

Examination methods of nose

Content

1. Sight, palpation
2. Rhinoscopy
3. Examination of nose patency
4. Examination of mucocilliary activity
5. Examination of olfaction
6. Examination of paranasal sinuses

Sight, palpation

We assess: nose configuration, line of nasal dorsum, possible deviations, crepitations, emphysema, skin color, exanthema, hemangioms, pigment naevi, ulcers or other pathology. We have to be aware of nose size and development deviations

Rhinoscopy

- Anterior rhinoscopy: with help of Hartmann´s mirror we can assess nasal entrance, septum, mucosa (pink, reddish, livid), secretion (clear, mucous, turbid, suppurating, bloody, with

crusts), configuration of lower and middle concha. Upper concha is inapproachable by direct sight.

- Middle rhinoscopy: sooner by Killian´s mirror (after topical anaesthesia and anemization of mucosa). Now it is used endoscopy of nasal cavity
- Posterior rhinoscopy: indirect examination of posterior parts of nasal cavity, choans, nasopharynx. It is done by mirror or now by endoscopy.

Examination of nose patency

- By Glatzel´s plate for guidance
- Insertion of sound or catheter into mesopharynx through nasal cavity (exclusion of choanal atresia in newborns)
- Rhinomanometry – measures intranasal pressure during inspiration and expiration
- Acoustic rhinometry – ultrasound based examination
- Endoscopy

Examination of mucocilliary activity

It s examined by saccharine test – after saccharine application on mucosa of nasal entrance patient senses sweet taste in throat after 15-20 minutes (time of substance transfer by mucocilliary transport into choans and onto taste caliculi.

Examination of olfaction

Olfaction disorders are divided into quantitative (anosmia, hyposmia) and qualitative (parosmia, phantosmia, specific anosmia)

- Quantitative (subjective) olfactometry – patient inhales different smells and odors and determinates them. Used substances are vanilla, coffee, lemon,…
- Qualitative olfactometry – according to Bornstein with help of 3 groups of substances:
    - Irritating only n.olfactorius (stearine, soap, camphoric spirit, lavender oil)
    - Stimulating n.V (menthol, ammonia)
    - Caused sense of taste (chloroform, pyridine)

Examination of paranasal sinuses

- X-ray examination – in frontal, semiaxial, axial and lateral projection. Conmeatusional X-ray gives information only about massive pathological mucosal changes (cysts, hyperplasia, polyps)
- CT (coronary projection – in frontal plane, axial projection – in transversal plane, possibly reconstruction) – displays mainly bones
- MRI displays very well soft tissues
- Frankel´s test – anemization of nasal mucosa with tampon with adrenalin or sanorin (5-10 min.) – leads to decongestion of mucosa and loose paranasal sinuses entrances open (presence of suppurating or serous-suppurating secretion under middle concha marks paranasal sinuses inflammation)

- Puncture of maxillary sinus – is indicated in case of cavity empyema. Needle is guided through lower nasal meatus, it goes towards lateral canthus
- Sinusoscopy – endoscopic examination. Maxillary sinus is examined through hole in maxilla created by trocar via fossa canina (caution – risk of teeth germs damage in young children) or lower nasal meatus. Frontal sinus is examined after communication creation in medial edge of eyebrow (Beck´s puncture). Sphenoidal sinus is examined through nasal cavity.

Congenital anomalies of nose

Content

1. Nose agenesis
2. Double nose
3. Proboscic lateralis
4. Medial or lateral rhinoschisis
5. Dermoid cyst
6. Stenosis and atresia of nasal entrance
7. Choanal atresia
8. Meningocele, encephalocele

Nose agenesis

Usually is developed only one nasal cavity, even sinuses are missing on afflicted side

Therapy: surgical in cooperation with plastic surgeon

Double nose

There are 2 noses with 4 nasal entrances

Therapy: surgical in cooperation with plastic surgeon

Proboscic lateralis

Creation of dermal and muscle protuberance in the shape of proboscis, which is located in the inner canthus. It is blind and usually contains sebaceous glands and cartilage.

Therapy: surgical in cooperation with plastic surgeon

Medial or lateral rhinoschisis

Left and right half of face is joined together during embryogenesis.

- Medial rhinoschisis is often combined with cheiloschisis. Incomplete rhinoschisis express itself as a: fissure of the apex nasi, medial fissure of nasal top wall, double nasal septum, fistulae nasi
- Lateral rhinoschisis is rare – it afflicts ala nasi, lateral wall of nose. They are rarely combined with eye or ear abnormalities

Therapy: surgical according to extent of rhinoschisis during first year of age

Dermoid cyst

If slot between frontal prominence and nasal bones is not closed, it is formed dermoid cyst. It is usually located in glabela area and in upper part of dorsum nasi or on nasal septum. It contains yellow pulpy substance, sometimes even with hair. Connection with intracranial space is possible.

Therapy: surgical, operation timing depends on size and possible inflammatory complications

Stenosis and atresia of nasal entrance

Unilateral or bilateral. Stenosis (nasal entrance narrower than 4mm) of nasal entrance is often found in patients with cheiloschisis.

Therapy: surgical in cooperation with plastic surgeon

Choanal atresia

Develops during 5.embryonal week, when oropharyngeal membrane didn´t rupture. Incidence is 1:5000, more frequently in girls, hereditary incidence is rare). It is often combined with other congenital anomalies (perceptive hearing loss, eye defects, polydactylia, etc.). Can be unilateral or bilateral, bone or cartilaginous.

Symptoms: depends on type of affliction. Bilateral atresia causes asphyxia immediately after birth. Newborn gets better during crying (open mouth) and gets worse during feeding (worsening of breathing). Unilateral atresia stays hidden until later age, it causes paranasal sinuse inflammations and rhinocleisis.

Diagnostics: ventilation test, sounding with nasogastric sound, endoscopy, CT, MRI

Therapy: bilateral congenital atresia demands urgent surgical intervention by endonasal endoscopic technique

Meningocele, encephalocele

It is prolapse of velamentum or brain during intrauterine development around 20.day. It manifests around 1.year of age. We divide them into outer and inner. Outer ones deform nasal area and orbits, inner ones go through lamina cribriformis into nose. They look like soft, glazy and sometimes pulsating formation.

Symptoms: worsen nose ventilation, liquorrhea (risk of intracranial infection)

Diagnostics: CT, endoscopy, beta-2-transferrine for liquor prove

Differential diagnostics: nasal polyps, tumor

Therapy: surgical, ATB

Diseases of external nose

Content

Seborrhoea nasi

Excretion disorder of sebaceous glands

Therapy: dermatological

Rosacea

Blood circulation defect – skin is red, number of teleangiactasies is increasing. Etiology is unclear. Known is influence of alcohol, coffee and hormonal changes.

Therapy: dermatological

Rhinophyma

Based on long lasting rosacea (massive thickening of skin on apex and alae nasi)

Therapy: surgical

Herpes simplex and zoster

It is caused by viral infection and cause circumscribed erythema, later vesicles and crusts. Neuralgic pain in n.V area can be present as well.

Therapy: symptomatic, in case of recurrence in combination with general therapy (Herpesin, Zovirax).

Erysipel

Infection disease of skin caused by Streptoccocus. It looks like painful, circumscribed red spot.

Therapy: penicillin, ATB with wide spectrum

Eczema of the nasal entrance

is developing after skin irritationwith pathological secretion during acute and chronic rhinosinusitis, in case of foreign body in nasal cavity or paranasal sinuses, physical or chemical agents or bad hygienic habits.

Therapy: removing of primary cause, chamomile compress, Jarish solution, unguents and oil softening, in case of dry eczema even corticosteroid unguents.

Foliculitis vestibule

Staphylococcal infection of hair follicle. It causes swelling and erythema with strong pain.

Furunculus nasi

It is disease with possible life threatening complications. It is staphylococcal infection of hair follicle or sebaceous gland. Infection isn´t only in the follicle area, but spreads into surrounding tissue and can cause flegmona or abscess. Disease is often followed by headache, strong pain, fever, regional lymphatic nodes swelling. Most sever complication is thrombosis of cavernous sinus, followed by sepsis and meningitis (infection spreads through venous system, which has no valves: v.facialis – v.angularis – v.ophthalmica).

Therapy: ATB against Staphylococci and drainage of suppurating focus. Removing by pressure is strictly forbidden!

Diseases of the nasal cavity and paranasal sinuses

Content

Rhinosinusitis

Terminology:

- Rhinosinusitis: inflammation of nasal mucosa and paranasal sinuses. Nasal cavity and paranasal sinuses form functional and anatomical unit. If natural orficies of paranasal sinuses are patent, inflammation afflicts mucosa of nasal cavity as well as mucosa of paranasal sinuses.
- Rhinitis: inflammation of nasal mucosa. Inflammation of nasal cavity without paranasal sinuses is possible only when natural orifices of paranasal sinuses are impassable. In praxis rhinitis means usually viral infection of nose and paranasal sinuses.
- Sinusitis: inflammation of paranasal sinuses mucosa. Isolated inflammation of paranasal sinuses is possible only when natural orifices of paranasal sinuses are impassable. In praxis sinusitis usually means bacterial or mycotic infection of nasal cavity and paranasal sinuses. Most frequently afflicted sinus in adults is maxillary, next is ethmoidal and frontal. Most frequent in children is ethmoidal inflammation. Inflammation can afflict only one sinus – monosinusitis, or more sinuses together – polysinusitis, or all sinuses – pansinusitis. If frontal group of paranasal sinuses is afflicted it causes pain of face, forehead or nose base. If posterior group is afflicted it causes pain in back of the neck. Bolesti is increased during cough and forward bend. Children can have different symptoms - local symptoms are not expressed, but general problems are bigger.

Definition:

recurrent 3 of followed symptoms: nose discharge, nose obstruction, sneezing, itching, cough.

Classification:

- Allergic:
    - Seasonal
    - Perennial
- Infectious
    - Acute (viral, bacterial)
    - Chronic (bacterial, mycotic)
- Others

Allergic rhinosinusitis

- Seasonal – watery secretion, sneezing and nose itching, it appears especially in the time of pollen season, eye symptoms are present as well. Typical allergens: pollen, grass, moulds
- Perennial – dominant is nose obstruction for whole year, eye affliction is uncommon. Typical allergens – acarids, dust, parasites, furs, cockroaches

Diagnostics: rhinoscopy: serous secretion, mucosal swelling, livid color of mucosa or erythema, nasl polyps. Allergologic tests.

Therapy: nasal corticosteroids, antihistaminics, surgery in case of polyps.

Acute rhinosinusitis

Etiology:

- viral (rhinovirus, adenovirus, RS virus, picornavirus)
- bacterial superinfection (s.pneumoniae, H.influenzae, S.aureus, M.catarrhalis, etc.) usually after 5-7 days from viral infection

Pathogenesis:

- inner causes: nose patency restriction, congenital or gained predisposition, infection focus in the nose or paranasal sinuses
- outer causes: climate, working environment

Infalmmation can develop by spreading from teeth, after long nasal intubation, in tampon insertion, after injury, by infected water in the swimming pool, in defects in nasal cavity. In sucklings are symptoms more sever, because fed baby has problems with swallowing because of nose breathing restriction.

Diagnostics: rhinoscopy – serous secretion in viral infections (changes into mucous), suppurating secretion in bacterial superinfection, erythema and mucosal swelling.

Therapy:

- nasal corticosteroids (don´t have systemic effects)
- antihistaminics: 1.generation (promethazin, dithiaden) – suitable in young children (sedation), 2.generation (Claritine, cetirizine) – do not have sedative effect
- nasal decongestion drops (contain antihistaminics and pseudoefedrine derivates – Clarinase, Disophrol)
- anemization nasal drops: better distribution if spray is used
- Salt solutions, Vincentka: suitable for after-treatment, in children and as a premeatusion
- ATB – in bacterial superinfection

Chronic rhinosinusitis

Definition: symptoms last at least 12 weeks, or 6 emeatuss of acute rhinosinusitis per year in children or 4 in adults, or presence of permanent changes on CT. Inflammatory chronic mucosal hyperplasia can be cause of cysts and polyps

Etiology: bacterial or mycotic

- Rhinosinusitis chronica atrophica – mucosal congestion, hyposmia, crusts, stench from nose. We divide: primary atrophic rhinosinusitis (ozaena) and secondary atrophic rhinosinusitis (after radiotherapy, surgery or injury)

Therapy: Vincentka, mucosal moistening, ATB, surgery

- Rhinosinusitis chronic simplex – the most simple form with increased nose secretion and temporary nose obstruction (reversible hypertrophy). It caused by conchal congestion.

Diagnostics: rhinoscopy: erythema and sweeling of mucosa, dense secretion. Mucosal swelling disappears after vasoconstriction therapy. Imaging methods (CT) is indicated before surgical intervention.

Therapy: medication (corticosteroids, antihistaminics), if it is unsuccessful – surgery (FESS)

- Rhinosinusitis chronica hypertrophica – permanently worsened patency of nasal cavities (irreversible hypertrophy), dense mucous secretion, hyposmia

Diagnostics: rhinoscopy: erythema and swelling of mucosa, desnse secretion. After vasoconstriction therapy we can see no effect. Imaging methods (CT) is indicated before surgical intervention.

Therapy: surgical (FESS, nasal mucosa ablation)

Other rhinosinusitis

- NARES – non-allergic rhinitis with eosinophilia syndrome. Perennial flu with sneezing, itching and discharge
- Professional
- Hormonal – pregnancy, adolescence, climax, endocrinopathy (thyroid gland, hypophysis)
- Induced by medication – sanorin, contraceptives, reserpine, chlorpromazine
- Psychic etiology – stress, sexual arousal – influenced by autonomous stimulation
- Alimentary – food, conservation agents, colors
- Idiopathic – (vasomotoric rhinitis) is nasal hyperactivity on non-specific trigger factors (warmth, cold)

Specific rhinosinusitis

- Rhinoscleroma of nose

It is chronic granulomatous inflammation of nasal mucosa caused by Klebsiella rhinoscleromatis. Infection enters organism via nose into airways. Scleroma is formed at first in nose and from there it is spreading into pharynx, trachea, larynx and lower respiratory tract. It can afflict even nose and face skin.

Therapy: ATB, surgery

- Rhinitis gonorrhoica neonatorum

It is caused by gonococci infection during birth. It is severe form of suppurating necrotic rhinosinusitis with ostitis of nasal skeleton, often with lethal end.

Therapy: ATB

- Rhinitis syphilitica neonatorum

It manifests in 3.week of life by stench secretion from nose, ulcer and fissures formation in nose entrance, lymphatic nodes enlargement. Morfologic changes in oral cavity are often present.

Therapy: ATB

Nasal polyps and cysts

Developing on base of chronic inflammation or allergy.

- Polyps are mucosal duplications – pedunculated formations formed by edematous bindweb and covered by gray mucosa. Meningocele, tumor or chronic foreign body must be excluded.

Antrochoanal polyp grows from maxillary sinus. Thanks to mucocilliary transport it grows into choans.

Multiple polyps grow from ethmoids.

- Thin-walled cysts are usually in maxillary sinus, they are filled with serous or mucous content. Exit form mucous gland is closed. Pressure of bigger cysts on cavity wall causes headache, but doesn´t cause any nose symptoms. Diagnostics – with X-ray in semiaxial projection.

Therapy: surgical (sinoscopy, polypectomy, FESS), corticosteroids

Disease of septum nasi

Deviatio septi nasi

Appears separately or in combination with deformities of whole nose. Can be congenital or can develop in adolescence as a result of asymmetric growth of cartilaginous and bone skeleton of the face or as a result of injury.

Symptoms: difficult unilateral nose breathing, compensatory hypertrophy of lower concha on other side, snoring, sleep-apnoe syndrome, olfaction disorders. Risk of inflammations is increased because of bad ventilation.

Therapy: septoplasty after end of growth

Perforatio septi nasi

Develops after septum operation, chemocaustics on both sides of nasal septum, in rhinitis sicca anterior, after abscess of septum, in specific inflammations of septum (TBC, syphilis), after cocaine or other addictive substances application, during work with chemicals. Septum perforation can be sign of tumor or other serious disease.

Symptoms: increased crusts formation, whistling murmur during nose breathing

Therapy: surgical (transposition of surrounding mucosa, implantation of autologous cartilage, prosthesis)

Bleeding polyp of septum

Can be source of recurrent epistaxes, therapy is surgical.

Synechia

Develops most frequently after injury, operation in nasal cavity, mucosa burn. We can see fibrous and mucosal bridges between septum and lateral wall of nasal cavity during anterior rhinoscopy.

Symptom: depends on localization and extent – cause mucus stagnation, difficult nose breathing s

Therapy: surgical – synechia disruption with local treatment (fat tampons) into epithelization of the mucosa.

Epistaxis

Develops from local as well as from general causes. It often develops in patients with hypertension, hematologic disease, tumors of nasal cavity, paranasal sinuses and epipharynx, venectasia of septum. Epistaxis follows nose injury or damage of nasal mucosa (foreign body, repeated suction). Most frequent is bleeding from locus Kiesellbachi – venous plexus in anterior part of nasal septum. Woodruf´s plexus is located on the posterior part of lower concha.

Diagnostics: rhinoscopy, nose endoscopy, hematologic and internal examination

Therapy: according to extent and bleeding localization.

- Firs aid: - head must be bent forward, everything must be blown out of nose. Press alae nasi into septum, cold compress, anemization
- In case of recurrent bleeding use chemocaustics (silver nitrate, chromic acid, hemostyptics, electrocaustics)
- In case of stronger bleeding:
    - Frontal package: by absorbable material (Gelaspon, Spongostan) or fat package. If frontal package is in the nose more than 48 hours, ATB are necessary as a premeatusion of infection.
    - Posterior package: tampon insertion into nasopharynx in case of bleeding localized dorsally in nasal cavity or nasopharynx. It is done by thin catheter inserted into nose. Catheter is pulled out from mouth and tampon is fixed on it. By pulling nasal end of catheter the tampon is fixed in nasopharynx, fibres are fixed to the face. Anterior package usually follows. ATB are necessary.

PCD – primary cilliary dyskinesis

Genetically conditional autosomal recessive disease. Cilia movement of cilliary epithelium is insufficient and uncoordinated – patients have problems caused by increased mucus production and stagnation in airways.

Symptoms: persistent suppurating rhinosinusitis, recurrent otitis, chronic cough, atelectatic focuses, chronic bronchitis. Majority of boys suffer from sterility in adult as a result of structural defect of sperms. About 50% of patients with PCD have dextrocardia (Kartagener´s syndrome – recurrent sinusitis, bronchiectaziae, dextrocardia)

Diagnostics: electron microscope examination of nasal mucosa, saccharide test or isotope examination.

Therapy: symptomatic – early ATB therapy of lower airways inflammation, mucolytics, climate therapy

Postnasal drip (sinobronchial syndrome)

Secretion from upper airways flows during rhinosinusitis into lower airways and causes bronchitis.

Symptoms: persistent cough, headache

Diagnostics: chronic rhinosinusitis during bronchtitic auscultation finding

Therapy: rhinosinusitis therapy, ATB

Cystic fibrosis

Symptoms: bronchial obstruction by viscous mucus, rhinosinustis, nasal polyps.

Diagnostics: endoscopy, electron microscope examination of mucosa

Wegener´s granulomatosis

Vasculitis combined with granulomatosis on autoimmune base.

Classification:

1. grade – ulcers in nasal cavity
2. grade – multifocal affliction with systemic symptoms
3. grade – systemic affliction with significant symptoms and multiorgans failure

Symptoms: nose obstruction with serosanquinolent secretion, crusts in nasal cavity, atrophic rhinitis, increased temperature, pain in middle part of face. In later stage are present infiltrates in nasal cavity and lungs, in 3.stage fever, hemoptysis, renal insufficiency.

Therapy: corticosteroids with cyclophosphamide, cotrimoxazolum, reconstruction of external nose (rhinoplasty).

Tumors of nose in children

Parameningeal rhabdomyosarcoma

Most frequent soft tissue malignant tumor of head and neck in children (40% of all RMS)

Symptoms: painless swelling of paranasal sinuses, orbits, nasopharynx, fossa pterygopalatina and fossa infratemporalis. Symptoms result from damage of surrounding tissues. Nose obstruction, epistaxis, proptosis, OMS, head nerves disorders. Destruction of anterior part of skull base can rarely cause pain. Almost all patients have metastases in the time of diagnostics.

Diagnosis: CT, MRI, biopsy, bone marrow, X-ray of bones, examination of liver, spleen, liquor, Ca, P, other blood tests.

Therapy: chemotherapy – vincristine, actinomycine, cyclophosphamide. Radiotherapy – main complications are face growth defects and X-ray induced tumors. Surgery – diagnostics from biopsy, resection of residual tumor after 12 months.

See also: Rhabdomyosarcoma in Tumors and expansive processes in children.

Angiofibroma

It is found only in adolescents – boys. It grows from foramen sphenopalatinum (upper edge) towards nasopharynx, through choans into nasal cavity, fossa infratemporalis. It grows to the paranasal sinuses, orbits and middle excavation. Tumor is usually white, spherical, nodulated, uncoated and covered by mucosa with big submucous vessels. Histologically it is formed from vascular and stromal components. Vessels are regularly placed and miss contractive elements (risk of strong bleeding, which can be stopped only with difficulties)

Symptoms: nose obstruction, epistaxis, rhinophonia causa, swelling of face, proptosis, diplopia, vision disorders, local bone destruction.Recurrences are common after operation.

Staging:

- 1A nose and nasopharynx
- 1B paranasal sinuses
- 2A foramen spenopalatinum
- 2B fossa pterygopalatina
- 2C fossa infratemporalis
- 3A intracranium

Diagnostics: nose endoscopy, CT, MRI, angiography, excision is contraindicated – risk of great bleeding

Therapy: surgery (preoperative embolization of tumor vessels during angiography), radiotherapy in case of intracranial spreading – it can cause X-ray induced tumor in irradiated area. Surgical approaches: most frequently endonasal surgery, transpalatinal, lateral rhinotomy, midfacial degloving. Controlled hypotension during operation.

Carcinoma of nasopharynx

Probably most frequent wrongly diagnosed tumor of head and neck. It includes 0,25% of all tumors in North America, but 18% in China (antigens HLA-A2, HLA-B-sin2 loci). Risk is increased by eating salty fishes (nitrosamine), exposure to tobacco smoke and dust, chronic infections of nose and paranasal sinuses, EBV, bad hygiene and insufficient ventilation. It occurs in any age, middle age is 51 years. It includes 1/3 of tumors in nasopharynx in children, without difference between genders.

Symptoms: metastases in lymphatic nodes, hearing loss, nasal obstruction and secretion, epistaxis, headache, neuropathy – especially growing into foramen lacerum.

Diagnostics: epipharyngoscopy, CT, MRI, biopsy, immunology (antibodies against EBV)

Therapy: radiotherapy 6.500 – 7.000 cGy + prophylactic irradiation of neck lymphatic nodes, neck dissection, resection of residual or recurrent tumor. In case of dissemination is used adjuvant chemotherapy. Five year survival in children is 40%.

Nose injuries

Injuries of face are very common. They develop during birth in suckling in case of fall on head, during development of movement abilities of child, in older children during sports etc. They can be irrelevant (surface scratches, hematoms), but also very serious, where eye brain and jaws can be damaged and even cosmetic impact is important.

Content

Injury of facial soft tissues

Surface injuries of nose are relatively often, contusion or laceration injuries combined with fractures of nasal bones and nasal septum

Therapy: suture, ATB

Fracture of the nose

Open or closed

Symptoms: nose deformation, bleeding, impaired nose ventilation, swelling, hematoma, pain

Diagnostics: sight, palpation (crepitations in case of fracture), rhinoscopy, X-ray

Therapy: cold compress, nasal drops, suture of skin, anterior package. Reposition of nasal bones should be made up to 7 days from injury and in children in addition in general anaesthesia.

Hematoma and abscess of septum

Develop during blunt punch, so that blood flows between perichondrium and mucosa of nasal septum. It is usually bilateral. In case of einfection develops abscess. Risk of this disease is in possible development of meningitis, sepsis and tromboflebitis of cavernous sinus (via v.angularis and v.ophthalmica).

Symptoms: difficult nose breathing, soft bulge of septum with fluctuation

Diagnostics: rhinoscopy

Therapy: incision, drainage, anterior package, ATB

Fractures of facial skeleton

Le Fort classification

- Le Fort I – abruption of maxillary alveolar processus (swelling of lower parts of face, occlusion disorders, pathological mobility of teeth arch)
- Le Fort II – fracture line goes from nasal base to the foramen infraorbitale, dorsally on tuber maxillae and to processus pterygoideus (swelling, hematoma, epistaxis, pathological occlusion, step on lower edge of orbit)
- Le Fort III – separation of facial skeleton from skull base – craniofacial abruption (bleeding from nose and mouth, hematoma, concave deformation of face, pathological occlusion, liquorrhea, anosmia, eye complications)
- Therapy: ATB, stomatosurgery. Operation is indicated after stabilization of state.

Other fractures

- Isolated fracture of processus zygomaticus – concave deformation and painful movement of temporomandibular joint
- Injury of temporomandibular joint – luxation – occlusion disorders
- Fracture of zygomatic complex – asymmetry of face, enophtalmus, pain of temporomandibular joint, eyelids hematoma, diplopia, hypestesia of 2.branch of n.V
- Isolated fracture of orbit base (blow out) – by blunt punch to the eye (e.g. tennis ball) – lower wall of orbit is broken, because the wall is architectonically most thin.

Symptoms: diplopia, restricted movement of eyeball (caused by eyeball muscles incarceration), eyelid hematoma, enophtalmus, disorders of n.V

Diagnostics: CT, eye examination

Therapy: ATB, surgery in case of eyeball movement restriction

- Fractures of anterior wall of frontal sinus – impressions are caused by fragments dislocation into cavity

Diagnostics: CT, eye examination

Therapy: ATB, surgery with fixation and elevation of fragments and with control of sinuses exits

- Frontonasoethmoidal fractures – if force acts in middle of upper etage. It causes fracture of superciliary ridge, medial wall of ethmoids, nasal bones or lamina cribrosa. Liquorrhea must be excluded. Damage of 1.branch of n.V and lacrimal pathways.

Diagnostics: CT, eye examination

Therapy: premeatusion of infection and brain edema (ATB, corticosteroids), surgery – close dura mater defect and stop bleeding

Foreign bodies in the nose

More often in right side (majority of right-handed) – peas, beans, stones, beads, paper, toys, polystyrene, etc. In case of injury or bullet wound foreign body can get to paranasal sinuses. Parts of food can get to nasal cavity during sneezing or vomiting

Symptoms: difficult ventilation, unilateral nose secretion, in case of chronic foreign body is present unilateral nose and paranasal sinuses inflammation.

Diagnostics: rhinoscopy, examination of nose patency, X-ray

Therapy: removing by hook, stuck bodies have to be removed in general anaesthesia. Tweezer is absolutely unsuitable tool.

Inflammatory complications of nasal diseases

Content

Local complications

Empyema of paranasal sinus

Usually in maxillary sinus during obstruction of natural orifices

Symptoms: pain (increased if patient bend forward), swelling and erythema of face

Diagnostics: X-ray semiaxial projection, CT, Frankel´s test, rhinoscopy

Therapy: ATB, nasal corticosteroids, antihistaminics, puncture, sinoscopy, supraturbinal antrostomy

Osteomyelitis

Symptoms: swelling above afflicted bone, swelling of eyelid, pain, fever

Diagnostics: rhinoscopy, CT

Therapy: surgical removal of afflicted bone, long term treatment with ATB

Mucocele, mucopyocele

Are rare in children. It afflicts especially frontal sinuses. It develops in case of inflammation, injury or surgery, if opening of sinus is permanently closed. Result is long lasting mucus and mucopus retention inside cavity, which causes pressure increase and bone deformation. According to content we divide them into mucocele (mucus), pyocele (pus), mucopyocele, hydrocele (watery content).

Symptoms: bulge near the inner canthus or on forehead, eyeball dislocation if mucocele causes pressure on orbital wall

Diagnostics: X-ray, CT. In differential diagnostics is necessary to differentiate meningocele and dacryocystitis.

Therapy: surgical

Orbit complications

Are most frequent. Cause is usually ethmoiditis and less often inflammation of frontal, maxillary or sphenoidal sinus or odontogenous inflammations. Inflammations of orbits can develop after injury or inflammation of eyelids or conjunctives, after insect bite, etc.

Classification:

Important is relation to so called orbital septum (periorbit + tarsal plates of eyelids)

- Chandler (1970):

1. Inflammatory edema – inflammation (flegmona) localized between orbital septum and bone wall of orbit
2. Orbital cellulitis – inflammation (flegmona) in orbit (spreads through orbital septum)
3. Subperiostal abscess – inflammation (abscess) in orbit (spreads through orbital septum)
4. Orbital abscess – inflammation (Abscess) localized in orbit (spreads through orbital septum)
5. Thrombophlebitis of cavernous sinus – most frequent complication of orbital inflammation

- Moloney (1987):

1. Epiperiorbital flegmona – same as 1. according to Chandler
2. Epiperiorbital abscess – same as 3. according to Chandler
3. Orbital flegmona - same as 2. according to Chandler
4. Orbital abscess

Symptoms: erythema and eyelids swelling, especially in the medial parts, chemosis of conjunctives, protrusion, deviation, dislocation and eyeball movement disorders, function disorders of II-VI head nerves, impaired sight or even sight loss, ptosis of upper eyelid, pain, panophthalmia

Diagnostics: CT, MRI, ENT and eye examination, neurology

Therapy: conservative in case of flegmona – ATB, therapy of rhinosinusits. Surgical if patient doesn´t get better in 48 hours or in case of abscess. Surgical approach is endonasal, external or combination of both.

Intracranial complications

- Meningitis
- Abscesses: epidural, subdural, brain (headache, nausea, bradycardia)
- Trombophlebitis of intracranial sinuses: sinus cavernosus (clinical symptoms are same as in case of bilateral orbit inflammation), sinus sagittalis superior

Diagnostics: CT, MRI

Therapy: ATB, surgery (neurosurgical craniotomy and abscess exstirpation), hematological treatment (thrombolysis in case of thrombosis of sinuses)

Distant complications

Infection focus develops during chronic inflammation of paranasal sinuses. Bacterial toxins spreads out of cavity into blood and cause allergic-hyperergic reactions. They can afflict heart, joints, kidneys, skin, eyes and vessels. Symptoms get worse if acute sinusitis is present.

Diagnostics: prove of chronic rhinosinusitis, bacteriology, allergology

Therapy: rhinosinusitis treatment

Basics of nose surgery

Endonasal surgery

Surgical interventions made by endoscopy with help of special instruments. Operation is less radical in comparison with classical surgery, however, it is technically more difficult. We try to spare physiological mechanisms (mucociliary transport). In presence it is important in therapy of inflammations, with limited value in tumors.

Terminology:

Used abbreviations: in literature are used those abbreviations and terms, their meaning is practically the same:

- FES (functional endonasal surgery)
- FESS (functional endonasal sinus surgery)
- EES (endoscopic endonasal surgery)

For simplicity reasons we will use in text abbreviation FES

Principles of FES:

FES states that mucosa of nose and paranasal sinuse has great regeneration ability, if those conditions are preserved:

- Sufficient ventilation
- Mucociliary transport

Most frequent surgical interventions:

- Sinusoscopy (sinoscopy)
- Supraturbinal antrostomy – enlargement of sinus maxillaris opening in middle nasal meatus. Anterior edge of hiatus semilunaris forms processus uncinatus ossis ethmoidalis. It is so called “unicnatectomy” (comment: os turbinale – lower nasal concha)
- Ethmoidectomia anterior – opening of anterior group of ethmoids
- Ethmooidectomia posterior - opening of anterior group of ethmoids
- Sphenoidectomia – opening and operation of sinus sphenoidalis

Septorhinoplasty

- Septoplasty: is made in case of nasal septum deformations and is usually done after end of growth (16 ears of age), earlier only exceptionally. Principle of operation is to remove deformed parts of cartilaginous or even bone skeleton of septum with returning part of cartilage.
- Rhinoplasty: from cosmetic reasons in case of external nose deformations

Classical surgery of nose and paranasal sinuses

In presence it is usually made in case of tumors, sometimes even for inflammation therapy

Most frequent operations:

Sinus maxillaries:

- Sec. Caldwell-Luc: resection of anterior wall of maxillary sinus (antrum Higmori – AH) form vestibulum oris superior, removing AH mucosa, communication formation into lower nasal meatus

Sinus frontalis:

- Sec. Beck: approach by opening formation in anterior wall of frontal sinus in medial ridge of eyebrow
- Sec. Jansen-Ritter: approach by resection of lower wall of frontal sinus
- Sec. Killian: approach by resection of lower and part of anterior walls of frontal sinus. Bone in supraciliary ridge is spared.
- Sec. Riedl: approach by resection of lower and anterior wall of frontal sinus. Cosmetic defect is inevitable.
- Osteoplastic operation: skin incision in upper edge of frontal bone with lifting off the skin lobe and anterior wall of frontal sinus and returning them back at the end of operation.

Ethmoids, sinus sphenoidalis, nasopharynx:

- Sec. Moore (lateral rhinotomy): approach skin incision on lateral wall of nose, resection of lateral bparts of nasal bones, ethmoidectomy
- Midfacial degloving: approach – incision in vestibulum oris sup., stalling skin of upper lip and nose cranially

Comments to embryology of oral cavity and pharynx

Development of face starts in 5.week of intrauterine evolvement. First element of oral cavity is stomodeum coated by ectoderma. Caudally is limited by lower processes of mandible arches which grow together in middle line into base of mandible. Cranially is wide frontal fornix, on sides are maxillary processes for upper jaw. Above frontal fornix are formed from ectoderma olfaction placods. Ectoderma proliferate and forms thick septum, which connects olfaction sockets with stomodeum. It gradually disintegrates and on its place is inserting mesenchyme, which forms primary palate. From ectodermal septum remain only two-layer oronasal membrane and this membrane

divide in dorsal part nasal vesicles from primitive oral cavity. In 38.-40.day cells of this membrane perish and primitive choans are formed.

Maxillary processes grow medially around lateral edges of nasal fornices and surround with medial nasal fornices primitive oral cavity. Biggest par of upper jaw and lip develops from mesenchyme with exception of interjaw segment, this one is formed from mesenchyme of medial nasal fornices and forms philtrum – part of jaw with incisors and primitive palate.

Primitive oral cavity is opening by primitive mouth entrance and nostrils it is placed directly on base skull. Bottom is formed by oropharyngeal membrane, which rupture at the and of 3.week and merge with ventral wall of pharynx. After this rupture oral cavity and pharynx are formed. Mesodermal palatal plates grow from processes for upper jaw in 6.week, they grow at first caudally with tongue descent have horizontal position. Plates meet in middle line, but grow together in 9.week. Uvula is formed from uvular processes which grow together. Hard palate is formed by ossification of maxilla and ossification of frontal part of palate. Dorsal part of palate stays as a soft palate and forms uvula.

With development ending of definitive palate is primitive oral cavity divided into definitive oral cavity and nasal cavity, which is dived from the beginning into left and right half. Fossa tonsillaris coated with entoderma is formed from 2.branchial evagination, which gets smaller. Entoderma in fossa tonsillaris proliferate into surrounding mesenchyme by processes which form bases of tonsillary crypts. Nodes of lymphatic tissue are formed along these crypts in 5.embryonic month. This development continues even after birth, mostly in first 6 months. Embryonic pharynx is meatusrodorsally flat funnel, extended in frontal plane, caudally narrowing into esophagus. On sides are ectodermal branchial evagiantions, against them are ectoderma-coated branchial sulci. Between ectoderma and entoderma stays thin layer of mesenchyme and from epithelium of branchial evaginations are formed branchiogenic organs.

Tongue is evolving in 4.week from several elements. Tuberculum impar evolves on inner face of ventral wall of primitive pharynx medially before foramen caecum. On its sides evolve paired lingual lateral tuberculi from mesenchyme of 1.brancial arch. Those 3 elements of tongue merge together. It grows cranially and frontally and form dorsum and apex linguae, which includes frontal 2/3 of tongue (inervation n.mandibularis, taste – chorda tympani). Radix linguae is formed from mesenchyme of 3.and 4.bracnhial arch (inervation n. glossopharyngeus). After birth have sucklings filled oral cavity with tongue. Tongue touches hard and soft palate and mucosa of faces and doesn´t have apex. This tight contact is very important during suction. With child growth and teeth development tongue gains same role as in adults.

Tonsilla pharyngea is formed on craniodorsal wall of pharynx in 6.month form entodermal epithelial plugs in form of mucosal plicae. Into those plicae penetrate lymphocytes. In 8.month is similarly formed tonsilla lingualis in the radix linguae.

Salivary glands are formed as solid cellular buttons, they grow from primitive oral cavity into surrounding mesenchyme. Large salivary glands are formed sooner than small ones in 2.embryonic month. Glandular differentiation is the same. Button formations lengthen, branching dichotonically on basics of ducts, their endings are differentiated into acins. From surrounding mesenchyme is formed fibrous intersticium and capsule of gland. First months after birth are salivary glands

relatively small with small salivary production. This is no problem for suckling. With teeth growth and food change glands are enlarging and produce sufficient amount of saliva.

Clinical anatomy of oral cavity and nasopharynx

Content

Oral cavity

It´s borders are formed by lips, base of oral cavity, hard and soft palate and plane of anterior palatal arches. It continues towards oropharynx through isthmus faucium (pharyngeal entrance, formed by palatal arches with tonsils, soft palate with uvula and base of tongue). In oral cavity is spinocellular uncornificational epithelium. In some parts is periosteum (processus alveolares, hard palate)In oral cavity are multiple subepithelial small salivary glands.

- Vestibulum oris is space between lips and face on one side and processus alveolares and teeth on the other side.
- Tongue – we divide it into: base, body, apex, dorsum and lateral edges. Under apex are plica sublingualis and caruncula sublingualis, where are located openings of submandibular and sublingual glands.
- Base of oral cavity – is formed especially by m.mylohyoideus
- Glandula parotis opens in face mucosa in the level of 2.molar tooth by ductus parotideus (Stenoni)
- Glandula submandibularis and sublungualis open under the tongue (Whartin´s duct), gl.sublingualis can have separated duct (Bartolini)

Vascularization

- Arterial:
    - A.carotis externa – a.lingualis, a.sublingualis, a.facialis
    - A.maxillaris – a.pharyngica superior, a.palatina inferior

- Venous:
    - Venous drainage to v.facialis through veins of same name, to v.jugularis interna throught pterygoideal plexus. Clinically important is connection through plexus pterygoideus to sinus cavernosus.
- Lymphatic
    - drainage through submental, submandibular nand parotidal nodes to the string of nodes along v.jugularis interna. Lymph drainage form base of oral cavity and tongue can be homolateral and even contralateral

Inervation

- Tongue
    - Motoric: n.hypoglossus
    - Sensitive: anterior part of n.lingualis, posterior part of n.vagus
    - Sensoric: n.glossopharyngeus, n.lingualis (posterior third), chorda tympani (anterior two thirds)
- Base of oral cavity
    - Motoric: n.mandibularis
    - Sensitive: n.trigeminus
- Muscles
    - Motoric: mastication muscles - n. mandibularis, mimic muscles – n.facialis
- Salivatory glands
    - Parasympathetic – n.VII – chorda tympani – ggl.submandibulare (gl.submandibularis, submentalis), n.IX – n.tympanicus – n.petrosus minor – ggl.oticum (gl.parotis)
    - Sympathetic – plexus carotideus
- Temporomandibular joint
    - N.mandibularis, n.auriculotemporalis

Pharynx

- Nasopharynx – superior part is formed by nasopharnyx vault, which is connected to skull base (sphenoidal bone). Posterior wall is formed by mucosa, submucous tissue and muscles laying on neck spondyls. In nasopharyngeal vault is lymphoepithelial tissue in children – tonsilla pharyngea. Lateral wall is formed by cartilage of torus tubarius, here is opened ET (connect nasopharynx with middle ear). Dorsally from torus tubarius is shallow fossa Rossenmuleri. Lateral and dorsal nasopharyngeal wall goes dorsally to lateral and dorsal oropharyngeal wall. It is formed by mucosa, submucous tissue and muscles of neck. Anerior wall of nasopharynx is formed by choans and soft palate. On dorsal wall of nasopharynx can be persistent bursa pharnygea. In the nasopharynx is cilliary epithelium, on the way to mesopharynx is changed into multi-layer flagstone epithelium.
- Oropharynx – it is oral part of pharynx – space formed by pharynx entrance (anterior) – istmius faucium, wall from mucosa, submucous tissue and muscles laying on neck spondyls (posterior). Lateral wall is near big vessels and nerves. Cranially goes to nasopharynx, caudally to hypopharynx -they are divided by upper edge of epiglottis.
- Hypopharynx – laryngeal part of pharynx, lateral and posterior wall have same structure as a walls of mesopharynx – space from upper edge of epiglottis to lower edge of ring cartilage.

On both sides along pharynx are mucosal evaginations – recessus piriformes, which are opened during swallowing. Mucosa is formed by spinocellular epithelium

Vascularizaiton

- Arterial – a.carotis externa
- Venous – v.facialis, v.jugularis interna, plexus pterygoideus
- Lymphatic – retropharyngeal nodes, deep neck nodes, paratracheal nodes

Waldeyer´s circle

It is formed by lymphatic tissue in pharynx and larynx:

- Tonsilla pharyngea – in nasopharyngeal vault
- Tonsillae tubariae (Gerlachi) – in fossa rosenmulleri
- Tonsillae palatinae – between palatal arches, they are divided from pharyngeal muscles by fibrous capsule
- Tonsilla lingualis – on the base of tongue
- Lymphatic tissue in the pharyngeal wall – on dorsal and lateral walls
- Lymphoepithelial tissue of meatusriculus laryngis Morgansky

Tonsils of Waldeyer´s circle are established embryonally, in childhood are enlarging as a result of developing immunity (especially in fist 6 years of age) as well as a result of recurrent infections. Lymphatic tissue of tonsils involves after adolescence.

Inervation

- Motoric: n.glossopharyngeus, n.hypoglossus, n.vagus
- Sensitizve: n.trigeminus, n.maxillaris, n.glossopharyngeus

Comments to anatomy and physiology of salivary glands

Salivary glands are located in maxillofacial area and their ducts open into oral cavity. We differentiate 3 big paired salivary glands (gl.parotis, submandibularis, sublingualis) and unpaired, small accessory glands (about 700) in mucosa of oral cavity and pharynx.

Glandula parotis

The biggest of the salivary glands lies in fossa retromandibularis, is located subcutaneously and have fibrous capsule (fascia parotidea), which is strongest on lateral side, medially is capsule not whole. Gland cranially reaches zygomatic arch, caudally goes about 1-2 cm under mandible angle to the m.sternocleidomastoideus, ventrally goes to edge of ascending branch of lower jaw, dorsally reaches meatus acusticus externus. Main duct – ductus parotideus Stenoni – is about 57 cm long, it goes from ventral part of gland, throught m.masseter, penetrates m.buccinator and facial mucosa. It opens in mucosa on the level of 2.molar tooth as a papilla parotidea. Clnically important is relation of gland to facial nerve. N.facialis leaves base skull through foramen stylomastoideum and enters glandular parenchyma as a short stem (0,5 - 1,5 cm in adults). He divides into 3 braches, which can have mutual anastomoses and form plexus parotideus. We differentiate terminal temporofrontal,

zygomaticobucal and cervical branching. Frontal branch and ramus marginalis mandibulae doesn´t have usually any anastomosis, otherwise course of branches is very variable. Medially from nerve fan (which divides gland into surface and deep part) lie branches of a.carotis externa providing gland with blood (a.maxillaris, a.retroauricularis,a.transversa faciei). Venous drainage is into v.jugularis interna. Lymphatic nodes (located intraglandulary and periglandulary) drain lymph through submandibular lymphatic nodes or directly into deep upper lymphatic nodes. Histologically is it serous gland, secretion cells form acins. N.facialis inervate mimic muscles of face and m.platysma.

Glandula submandibularis

IT is located in trigonum suibmandibulare, surrounded by m.bimeatuser, ligmantum stylomastoideum and mandible. Mai part of gland lies caudally from m.mylohyoideus, it is coverd by neck fascia. Its ductus submandibularis Whartoni is about 5-7 cm long, goes under mucosa of oral cavity base ventrally and opens near frenulum linguae in caruncula sublingualis. Near gland goes ramus marginalis mandibulae n.facialis (between upper part of gland and mandible), n-lindualis and n.hypoglossus, which must be spared during surgery on this gland. Vegetative inervation – n.lingualis. Gland is histologically mixed with serous and mucinous secretion cells.

Glandula sublingualis

Smallest paired salivary gland lies under mucosa of oral cavity base in plica sublingualis, dorsally from frenulum linguae, it touches anterior part of submandibular gland. Posterior group usually have separated ducts on carunculae sublinguales. Vegetative inervation – n.lingualis. Gland is histologically mucinous.

Small salivary glands

Are located in mucosa of oral cavity, density is variable, they are marked according to localization (e.g. gl.labiales, gl. buccales, gl.palatines, gllinguales posterior et laterals) We can find them in oral cavity mucosa, in inner side of lips, on palate, inside the mucosa of faces, pharynx, tongue, paranasal sinuses and larynx. The produce about 5-8% of all saliva, but in case of damage or removal of several big salivary glands are able to maintain sufficient humidity of mucosa. Small salivary glands are usually mucinous, serous are only Ebner´s glands around papills of tongue.

Function of the salivary glands

Saliva production is stimulated by physical, chemical and psychic factors. Salivary glands in suckling are small and produce only limited amount of saliva. With change of food the start to enlarge and produce bigger amount of saliva. After 24 hours is produced in adults about 1-1,5 litres of mixed saliva. Saliva is colorless or whitish viscid fluid composed from 99,5% of water and 0,5% of organic, anorganic and cellular substances. In saliva are excreted viruses in viremia, metals, microelements etc. Proportion of particular glands on saliva production is variable (biggest amount is formed in submandibular glands – over 50%). Saliva production can be influenced pharmacologically (e.g. pilocarpine).

- Xerostomia (sicca syndrome): dryness of oral cavity mucosa caused by insufficient saliva production – in case of dehydration, after irradiation, disease of salivary glands, central affliction of autonomous nerve system of salivary glands, in Sjórgen´s syndrome.
- Sialorrhoea – excessive saliva production – e.g. during teeth eruption, psychological factors
- Ptyalismus- increased saliva efflux (ptyalismus gravidarum) – can be pathological e.g. in case of neurologc diseases (epilepsy, morbus Parkinson)

Saliva function:

- Digestion – pestle food, moisten mucosa of oral cavity, starts digenstion with ptyaline, contains even lipolytic and proteolytic enzymes.
- Teeth protection – with help of enzymes and mechanically removes rests of food, with fluorine helps maintaining of teeth enamel, participate in teeth plaque formation.
- Taste – taste calyces
- Immunological defense – antiinfectious function – contains bactericide and bacteriostatic substances (lysozyme, properdine, immunoglobulins – especially IgA)
- Substances excretion from organism – I, Ca, Fe, Hg, Pb, As, alkaloids, halogens, coagulation factors, viruses (EBV, CMV, coxsackie, rubeola, poliomyelitis)

Examination methods of oral cavity and pharynx

Content

1. Nasopharynx examination
2. Oral cavity and mesopharynx examination
3. Hypoharynx examination
4. Salivary glands examination

Nasopharynx examination

- Posterior rhinoscopy – is made by small mirror and oral spatula. Mirror is guided behind soft palate. We see choans, torus tubarius, tonsilla pharyngea.
- Epipharyngoscopy – endoscopic examination through nasal or oral cavity
- Palpation –now in anaesthesia with finger thorugh oral cavity into nasopharynx
- Imaging methods – CT, MRI, X-ray (nasopharynx can be seen on lateral projection of skull), angiography

Oral cavity and mesopharynx examination

- Sight – lips, circumoral area, tongue, salivary glands openings, gingival, teeth. In mesopahrynx is assessed pharyngeal hilum, symmetry of arches, mobility of soft palate. We describe mucosa, its surface, color and roughness. During examination of tonsils we describe their size, symmetry (asymmetry is suspected from tumor), color and surface. Considerably increased tonsils can touch themselves in the middle line. Physiologically have tonsils smooth surface and pink color with jagged crypts. If we press on anterior palatal arch, healthy tonsil is medially luxed with ease, afflicted one stays in bed. Necessary for examination is artificial light and solid spatula.
- Palpation

- Imaging methods – USG, X-ray, CT, MRI
- Sialography – examination of salivary glands with contrast fluid

Hypopharynx examination

- Indirect hypopharyngoscopy – examination by laryngoscopic mirror. It is advantageous to pull out the tongue from mouth. We examine strctures of hypopharynx and larynx in reversed picture - base of tongue, epiglottical valecules, epiglottis, arytenoids tubers in dorsal part of laryngeal entrance, recessus piriformes laterally and dorsally which verge into esophagus. Recessus piriformes are physiologically without content, in case of esophagus impassability we can see saliva lakes.
- Direct hypopharyngoscopy – necessary is premedication and local mucosal anaesthesia, in children usually general anaesthesia and hospitalization. Metal tube is guided through oral cavity while head bending deeply backward. We see structures in direct picture.
- Imaging methods – X-ray, CT, MRI

Salivary glands examination

- History: general state of patient, clinical findings, epidemiological relationships, change and growth speed of formations, pain
- Findings according to age:
    - Neonates – often congenital lymphangioms, hemangioms
    - Scholl age – parotitis epidemica (mumps – even in adolescents), recurrent parotids (morbus Payen)
    - Middle age – adenoms, sialoadenoms. Number of malignant tumors is increased with age.
- Sight and palpation: asymmetry of face, edema, erythema, displacement of auricle, changes on skin, size and character of swelling, mobility of formation, function of n.facialis, sensitivity on pressure. In oral cavity: character and erythema of salivary glands openings, amount and character of saliva flowed out of salivary ducts during compression, concretion in Whartin´s duct.
- Imaging methods
    - USG - used to be the first imaging method
    - X-ray – can prove only contrast stone
    - Sialography – X-ray with contrast fluid application into salivary gland duct. It is made in two mutually perpendicular levels. Application of contrast is made by catheter inserted into duct. It is possible to make CT after contrast application.
    - Scintigraphy – salivary glands can accumulate particles of intravenously administered isotopes. Technecium 99 is used and its distribution in gland is recorded on special camera.
    - CT
    - MRI
- Biopsy: fine needle biopsy can give knowledge for surgical intervention indication.

Congenital defects of oral cavity and pharynx

Cleft defects

- Cheilognathopalatostaphylouvuloschisis – split of upper lip, upper jaw, palatal bone, soft palate and uvula

Split can be unilateral or bilateral, total or partial. It is genetically conditioned and treated surgically. Necessary is tight cooperation of plastic surgeon, ENT specialist and foniatrist.

Defects of tongue

- Split of tongue – rarely found, together with palatoschisis
- Frenulum linguae breve – movement restriction of tongue because of sublingual frenulum (suction in neonates, speech in older children). Therapy is surgical – frenulectomy.
- Macroglossia – in M.Down, hypothyreosis, acromegaly. We must exclude congenital tumors (hemangioma, vascular anomalies, neurofibromatosis)
- Accesory thyroid gland of tongue – it is more frequent in girls in case of not descending or not obliterating ductus thyreoglossus. Can be without symptoms, sometimes can be the accessory gland only fucnctional thyroid gland. Diagnostics: scintigraphy, endocrinological and ENT examination.

Adenoids and hypertrophy of palatal tonsils

Adenoids

Definition: patologick tonsilla pharyngea
Etiology: genetic predisposition, nutrition, immune system
Patophysiology: hypertrophy if lymphatic tissue restricts nose breathing and thats why can lead to airways infections. Obstruction of ET orifice decreases its function, which can lead to middle ear inflammation. Chronic inflammation of lymphatic tissue can cause recurrent local and distant infections.
Symptoms:
Caused by rectricted nose patency:

- mouth breathing
- snoring
- recurrent infections of airways and middle ear
- development disorder of facial skeleton
- facies adenoidea – face with open mouth and without nasolabial sulci
- enuresis nocturna – centrum of urination is stimulated during night due to hypoxia

Caused by chronic infection of lymphatic tissue:

- bacterial superinfection in viral infections of airways and middle ear
- distant (focal) infection: damaged tissues and organs by antigen-antibody complexes (joints, kidneys, heart, skin, eyes, ...)

Diagnostics:

- history: adenoid have all children (excluding those after adenotomy, however, recurrencies are possible)
- examination of nose and nasopharynx (endoscopy)

Therapy: adenotomy

Hypertrophia of palatal tonsils

Definiton: lymphatic tissue hypertrophy of palatal tonsils, which overreach palatal arches. Possible is coincidental chronic inflammation (tonsillitis chronica hypertrophica)
Etiology: genetic predispositions, nutrition, immune system, age (mostly in 6-7 ears of age)
Symptoms:

- OSAS (obstruction sleep apnea syndrome) causing apnoic pauses (due to relaxation of pharyngeal muscles), child repeatedly wake up and fall asleep
- Mouth breathing
- Snoring
- Damaged development of facial skeleton (gothic palate, oclusion disorder)
- Enuresis nocturna – centrum of urination is stimulated during night due to hypoxia
- distant (focal) infection: damaged tissues and organs by antigen-antibody complexes (joints, kidneys, heart, skin, eyes, ...)

Diagnostics: history, mesopharynx examination, nasopharynx examination, examination in sleep laboratory
Therapy: tonsillotomy in case of simple hypertrophy, tonsillectomy in case of coincidental chronic tonsillitis

Inflammations of oral cavity and pharynx

Content

3.1 Tuberculosis
3.2 Lues

Inflammations of oral cavity

Angulus infectiosus

Definition: fissures in labial angle

Etiology: bacterial or mycotic infection

Diagnostics: local finding, microbiological examination

Differential diagnostics: sideropenic anemia, decreased stamina of organism, diabetes mellitus in adults or lip carcinoma

Therapy: local therapy with silver nitrate 2-5%, ATB locally

Stomatitis aphtosa

Definition: vesicles with red rim (aphtae) on oral cavity or tongue mucosa. They break up and cause mucosal defect covered by fibrin film.

Etiology: isn´t known

Symptoms: elevated temperature or fever in first 2-3 days, child doesn´t want to eat

Diagnostics: local finding

Differential diagnostics: histiocytosis X, bowel parasites

Therapy: vitamins, analgetics, gentian violet, chamomile, accented hygiene of oral cavity, in recurrent or chronic forms ATB are recommended because of bacterial infection risk.

Soor

Definition: small or large coating on buccal mucosa, tongue, soft palate, arches and tonsils

Etiology: most frequently Candida

Pathogenesis: in suckling and children with depression of immune system, insufficient hygiene of oral cavity

Diagnostics: local finding

Therapy: gentian violet, chamomile, in more extended forms antimycotic therapy according to cultivation results

Herpes simplex

Definition: vesicles on lip or mucosa of oral cavity, crusts are forming

Etiology: virus herpes simplex

Pathogenesis: in case of immune system depression (fevers, infectious disease, exhaustion, etc), it last about 1-2 weeks. It is the most prevalent viral disease, up to 90% of population are carriers of virus.

Symptoms: pain

Diagnostics: local finding

Therapy: zinc unguent, Zovirax ung., orZovirax p.o. in case of extensive infection or if generalization menace.

Complications: herpetic meningitis

Herpes zoster

Definition: fast forming vesicles along nerves (e.g. branch of n.trigeminus), causes surface fibrous-epithelial defects

Etiology: virus herpes zoster

Symptoms: very painful disease, rare in children

Diagnostics: clinical finding – typical segmental course, or serologic examination

Therapy: Zovirax, vitamin B group, analgetics, ATB in case of bacterial superinfection

Noma oris

Definition: progressing inflammation – wet gangrene, starting in lip angle and spreading laterally, cause tissue necrosis

Pathogenesis: develops in children with severe malnutrition

Diagnostics: local finfing, general state of child

Therapy: ATB, surgical removal of necrosis, vitamin supplementation (especially B and C)

Abscess of tongue base

Etiopathogenesis

Develops after penetration of infected foreign body into tongue or by infection spreading from lingular tonsil, rare in children

Symptoms: restricted movement of tongue, strong pain with impossible food intake, fever

Diagnostics: history, local finding, USG

Therapy: surgical – incision and drainage of abscess, parenteral nutrition, wide spectrum parenteral ATB. It is life threatening disease.

Angina Ludowici

Definition: flegmona of oral cavity base

Etiopathogenesis: injury, foreign body, salivary glands infection

Symptoms: submental erythema, firm edema, pain

Diagnostics: history, local finding, USG

Therapy: surgical – incision, parenteral nutrition, wide spectrum parenteral ATB

Inflammations of pharynx

Acute inflammation of pharynx

Tonsillopharyngitis acuta

Definition: inflammation of mucosa and lymphatic tissue in pharynx

Etiology: respiratory viruses, possible bacterial superinfection

Course: they occur through whole year. It spreads usually air-borne, incubation time is 2-4 days.

Symptoms: Pain in throat, dysphagia, odynophagia, fever, flu, cough, swelling of regional lymphatic nodes

Diagnostics: msecopharynx examination, erythema and edema of mucosa and lymphatic tissue of tonsils

Therapy: analgetics, NSAIDs, ATB in bacterial superifnection

Tonsillitis acuta purulenta

Definition: acute inflammation afflicting lymphatic tissue around pharyngeal opening

Etiology: bacterial (streptococcus beta-hemolyticus, staphylococcus, pneumococcus, haemopillus). Scarlet angina – beta hemolytic streptococci

Pathogenesis: decreased constitution of organism (cold, other diseases)

Symptoms: throat pain, fever (above 38°C), swelling of regional lymphatic nodes (most frequently Wood´s nodes in mandible angle), dysphagia, odynophagia. In case of scarlet angina we can see maculopapulous exanthema in upper part of body, raspberry tongue (erythema and hyperplasia of papills), vesicles near nails on hands. Skin on fingers can desquamate around 8.day, if disease in not treated or treated too late.

Diagnostics:

- Lab tests: CRP, leucocytes, bacteriology, serology – scarlet exotoxin

- Mesopharynx examination: whithish coating of variable extent on palatal tonsils (tonsillitis lacunaris, follicularis, confluens)

Therapy: ATB (penicillin, cefalosporins, in case of allergy on previous ones macrolides). CAVE aminopenicillins – see mononucleosis. Scarlet fever – patient isolation.

Infectious mononucleosis

Etiology:

- Ebstein-Barr virus
- Cytomegalovirus (less severe course)

Pathogenesis: infection is transmitted via mucosal contact ("kissing disease") or air-borne. Often is collective infection. Incubation is 2-6 weeks.

Symptoms: usually asymptomatic, can be light flu, chronic fatigue syndrome. In more sever cases can be seen symptoms as in purulent tonsillitis with significant reactions of regional lymphatic nodes and possible hepatosplenomegaly.

Diagnostics:

- Mescopharynx examination, examination of lymphatic nodes in axillae, groins, size of spleen and liver
- Lab test:
    - leucocytosis with monocytes prevalence and cellular atypies
    - Paul-Bunnel test, antigen examination – EBV, CMV (IgM – acute disease, IgG – infection in the past), liver test – often are increased (AST, ALT, LD)

Therapy: symptomatic, liver diet, hepatoprotecive medication, tranquility (6 months without physical stress – rupture of spleen!). In case of secondary infection ATB (not aminopenicillins – cause toxoallergic reactions and liver damage). Corticosteroids only in the most sever forms.

Herpangina

Etiopathogenesis: coxsackie virus. Usually children of pre-school and school age in summer time. Incubation is 2-6 days.

Symptoms: fever with throat pain, loss of appetite, dysphagia, swelling of regional lymphatic nodes

Diagnostics: mesopharynx examination: on mucosa of pharyngeal hilum are vesicles with red rim, after rupture erosions develop.

Therapy: symptomatic: antipyretics, analgetics, local antiseptics, anaestetics, gentian violet. ATB only in case of secondary microbial infection.

Pharyngitis phlegmonosa

Etiopathogenesis: streptococcal infection after hypopharynx or esophagus injury by foreign body. Acute course with septic state, infection can spread into larynx and mediastinum.

Therapy: intensive parenteral application of wide spectrum ATB, parenteral nutrition until we assess extent of injury in swallowing pathways

Plaut-Vincent´s angina

Etiology: Bacillus fusiformis together with Spirocheta buccalis

Symptoms: throat pain, elevated temperature, swelling of regional lymphatic nodes

Diagnostics: mesopharynx examination: gray-white coating of triangle shape in upper pole of tonsil, unilateral affliction

Therapy: ATB

Chronic inflammations of pharynx

Tonsillitis chronic

Definition: chronic inflammation of lymphatic tissue of palatal or nasopharyngeal tonsil

Etiopathogenesis: S.pyogenes, often result of acute tonsillitis

Symptoms: often asymptomatic. Scratching, burning in neck, dysphagia, odynophagia, stench from mouth, swelling of regional lymphatic nodes, recurrent infections of respiratory airways (acute rhinosinusitism tonsillitits, etc.), otitis, metatonsillary complications (damage of distant tissues and organs: kidneys, joints, heart, eyes, skin, etc.)

Diagnostics:

- Mesopharynx examination: erythema of palatal arches, fixation of tonsils, hypertrophy and asymmetry of palatal tonsils, presence of plugs in tonsillary crypts
- Examination of nasopharynx: presence of plugs in nasopharyngeal tonsil, hypertrophy
- Lab tests: ASLO, bacteriology

Therapy:

- Adenotomy
- Tonsillectomy if patient has clinical problems or if complications occur

Pharyngitis chronica

Definition: chronic inflammation of pharynx

Pathogenesis: functional disorders of mucosa, chronically acting exogenous noxious agents, chronic disease, state after tonsillectomy. It is more often in adults.

Symptoms:

- Pharyngitis chronica simplex – dry pharyngeal mucosa or on the contrary erythema with secretion increase, usually without fever

- Pharyngitis chronica hypertrofica (granulomatosa) – thickness, swelling of pharyngeal mucosa with prominences of islets or strips of lymphatic tissue, production of sero-suppurating secretion, which often irritates and forces cough (especially in the morning), sense of foreign body in the throat
- Pharyngitis chronica atrophica (sicca) – in children only exceptionally, rather in older, especially in women. Posterior wall of pharynx is dry, flat with senses of dryness and burning in throat, which forces repeated swallowing, often with sense of foreign body. Laryngitis sicca nad atrophic rhinitis is often present as well.

Diagnostics: pharynx examination, bacteriology

Dif.dg.: specific pharyngitis, M.Sjógren, M.Plummer-Vinson

Therapy: symptomatic – fluids, inhalation, local antiseptics, antiflogistics, vitamins, immunostimulants, according to general state eventually ATB

Specific inflammations

Tuberculosis

Etiology: Mycobacterium tuberculosis, bovis

Pathogenesis: it is very rare in oropharynx. Primary complex develops after consummation of infected food.

Symptoms: ulcers on pharyngeal mucosa and tonsils with affliction of regional lymphatic nodes or sometimes with fistulae formation

Diagnostics: history, local finding, microbiology, serology, USG of lymphatic nodes, X-ray of chest, histology.

Therapy: antituberculotics, therapy is controlled by pneumologist

Lues (syphilis)

Etiology: Treponema pallidum

Pathogenesis: congenital or gained syphilis in suckling

Symptoms:

- Congenital lues:
    - early symptoms: pseudomembranes of pharynx and palatal arches, radial Parrot´sscars around mouth, syphilitic coryza, painless enlargement of regional lymphatic nodes
    - late symptoms: Hutchinson´s trias (barrel-like incisive teeth, parenchymatous keratitis, labyrintitis)
- Gained lues:
    - I.stage: tonsillitis with coating and lymphatic nodes swelling without fever

- II.stage: maculopapulous exanthema
- III.stage: gumma (destruction of palate and facial skeleton), lingua lobata – flat and grooved tongue

Diagnostics: history, local finding, serology (Bordet-Wasserman reaction)

Therapy: ATB, therapy is controlled by venerologist

Inflammatory complications of oral cavity and pharynx diseases

Content

1. Abscessus intratonsillaris
2. Phlegmona et abscessus peritonsillaris
3. Phlegmona et abscessus retropharyngealis
4. Phlegmona et abscessus parapharyngealis
5. Thrombophlebitis
6. Sepsis
7. Metatonsillary complications

Abscessus intratonsillaris

Is formed by several deep suppurating follicles of tonsil. It looks like yellow formation of variable size.

Therapy: ATB, incision, dilatation, tonsillectomy

Phlegmona et abscessus peritonsillaris

Definition: infection of peritonsillar space (between tonsil capsule and pharyngeal muscles) during acute or chronic tonsillitis

Etiopathogenesis: inflammation is spreading through tonsil capsule into surrounding space via fissures or along vessels. It is most frequent in 15-30 years of age. At first it has character of phlegmona, in several hours is formed abscess.

Classification:

- Supratonsillar phlegmona or abscess in upper pole of tonsil (99%)
- Infratonsillar phlegmona or abscess in lower pole of tonsil
- Retrotonsillar phlegmona or abscess near posterior palatal arch
- Symptoms: throat pain, dysphagia, odynophagia, fever, snuffing voice, difficult opening of mouth (trismus caused by spasms of chewing muscles), dyspnea in case of spreading into larynx (infra and retrotonsillar phlegmona or abscess)

Diagnostics:

Mesopharynx examination: usually unilateral affliction, bilateral is exceptionally rare

- Supratonsillar phlegmona or abscess: asymmetry of anterior palatal arches, bulge of arch on afflicted side

- Infratonsillar phlegmona or abscess: asymmetry of tongue base, bulge on afflicted side
- Retrotonsillar phlegmona or abscess: asymmetry of posterior palatal arches, bulge of arch on afflicted side

Therapy: ATB, in case of abscess surgery

- Supratonsillar absces :
    - Tonsillectomy á chaud (in the day, when diagnosis is determinate)
    - Tonsillectomy á tied (after treatment of acute state – after 1 week) with previous puncture, incision and abscess dilatation
    - Tonsillectomy á froid (after several weeks) with previous puncture, incision and abscess dilatation
- Infra and retrotonsillar abscess:
    - Tonsillectomy á chaud

Recurrences after paratonsillary abscess are in 10-30% and the only premeatusion is tonsillectomy.

Phlegmona or abscessus retropharyngealis

Definition: inflammation of retropharyngeal space between posterior part of oropharynx and paraevertebral fascia

Etiology: most often S.pyogenes

Pathogenesis: in suckling and toddlers is most frequent infection spreading from surroundings (infection of upper respiratory airways), in adults rather by injury of pharyngeal wall by foreign body

Symptoms: life threatening, fever, snuffing voice, odynophagia, pharyngeal stridor (in inspirium and exspirium), dyspnea, enlargement and pain of regional lymphatic nodes, paramedical bulge of posterior pharyngeal wall.

Diagnostics: history, local finding, clinical state, CT, lab tests (CRP, leucocytes, FW)

Therapy: incision and abscess drainage (transoral or external approach), parenteral ATB, general stabilization (secure airways)

Complications: intracranial infection, mediastinitis

Phlegmona et abscessus parapharyngealis

Definition: soft tissue infection of parapharyngeal space out of lateral wall of pharynx

Etiopathogenesis: in tonsillitis, infection of airways. Infection is spreading:

- Per continuitatem
- Via blood
- Via lymphatic system

Symptoms: swelling and edema of soft tissues on neck, erythema, fluctuation in abscess, fever, dyspahgia, odynophagia, dyspnea

Diagnostics: history, local finding, USG, CT, lab tests (CRP, leucocytes, FW)

Therapy: incision and drainage of phlegmona or abscess (transoral or external approach), parenteral ATB, general stabilization (secure airways)

Thrombophlebitis

Definition: inflammation of veins on neck with partial or total thrombus obstruction

Etiopathogenesis: during tonsillitis or respiratory infections – hemtogennous infection spreading. V.jugularis is afflicted most frequently. Affliction of v.facialis is described as a Lemier´s syndrome.

Symptoms: swelling and edema of soft tissues on neck, erythema, septic fever, strong pain along the vein

Diagnostics: history, local finding, USG, CT, lab tests (CRP, leucocytes, FW), microbiology

Therapy: anticoagulation with cooperation with hematologist (warfarine), ATB. Surgical intervention (resection of vein) if conservative treatment is unsuccessful. Infection source treatment (tonsillectomy, adenotomy)

Complications: intracranial infection, mediastinitis

Sepsis

Pathogenesis: bacteria or their toxins are present in the blood

- Sepsis tonsillogenes: septic focus is in palatal tonsil

Symptoms: septic fever, shake, weakness in acute tonsillitis

Diagnostics: history, local finding, leucocytes, CRP, positive cultivation from blood, sepsis

Therapy: tonsillectomy á chaud with administration of wide spectrum ATB

- Sepsis post angina: thrombophlebitis of neck veins after consolidation of suppurating angina

Symptoms: swelling and edema of soft tissues on neck, erythema, septic fever, strong pain along the vein

Diagnostics: history, local finding, leucocytes, CRP, positive cultivation from blood, sepsis, USG, CT

Therapy: anticoagulation with cooperation with hematologist (warfarine), ATB. Surgical intervention (resection of vein) if conservative treatment is unsuccessful. Infection source treatment (tonsillectomy, adenotomy)

Complications: intracranial infection, mediastinitis

Metatonsillar complications

Definition: affliction of distant tissues and organs (most frequently: joints, kidneys, heart, eyes, skin) in chronic or acute inflammations of lymphatic tissue of Waldeyer´s circle.

Etiology: beta-hemolytic Streptococcus

Pathogenesis: antibody formation against antigens from destroyed microbial cells. Complexes antigen-antibody are created.

Symptoms:

- Febris rheumatica – in children and adolescents. It appears after days or weeks after pharyngeal inflammation. Fever, joints pain, pathologic ECG.
- Nephritis – secondary streptococcal kidney disease. Appears 2-4 weeks after scarlet fever or streptococcal infection of tonsils.
- Affliction of heart, skin, eyes, etc.

Therapy: penicillin followed by premeatusive therapy by depot penicillin, tonsillectomy, adenotomy

Disease of salivary glands

Content

1. Injuries of salivary glands
2. Inflammations of salivary glands
    2.1 Parotitis epidemica (mumps)
    2.2 Cytomegalia of salivary glands
    2.3 Sialoadenitis acuta
    2.4 Parotitis recidivans (M.Payen)
    2.5 Sjógen´s syndrome
    2.6 Mikulicz syndrome
3. Sialolithiasis
4. Tumors of salivary glands
    4.1 Benign
    4.2 Malignant

Injuries of salivary glands

- Surface injury above salivary glands -suture in local anaesthesia
- Injury of gland duct – reconstruction in case on injury of main duct
- Salivary fistula –if main duct is intact, it usually closes itself. We restrict secretion by atropine. If it don´t close, we must surgically extirpate the fistula.

Inflammations of salivary glands

Parotitis epidemica (mumps)

Etiology: paramyxoviruses

Symptoms: swelling of mostly glandula parotis with swelling and erythema of its duct. Simultaneously can be afflicted pancreas, testes, ovaria, CNS. Hearing can be irreversibly damaged due to neurotropy of the virus (unilateral or bilateral deafness). After disease is lifetime immunity.

Diagnostics:

- History: incubation time is about 20 days
- Serology: direct prove of virus is possible only at the beginning of disease (saliva, urine, CSF)
- Amylase in blood and urine – maximum during 3.and 4.day.

Therapy: symptomatic – fluids, tranquility, analgetics, antipyretics, corticosteroids if orchitis is present

Complications: meningoencephalitis, hearing loss or deafness, orchitis, epididymitis

Cytomegalia of salivary glands

Etiology: cytomegalovirus (CMV)

Symptoms: possible kidneys, liver, lung and GIT affliction in case of generalized form in neonates and children till 2.year of age. In adults has non-characteristic symptoms of infection, noticeable is great fatigue without apparent cause. In children are present swellings of salivary glands with cyst formations, in severe cases hepatosplenomegaly with jaundice, thrombocytopenia, hemolytic anemia, chorioretinitis.

Diagnostics: history, local finding, general stat, serologic confirmation of antibodies

Therapy: symptomatic, immunoglobulins

Complications: psychomotoric and psychic retardation, affliction of written organs, death in neonates

Sialoadenitis acuta

Definition: bacterial inflammation of ascendant type from general and local causes.

Pathogenesis: onset in decreased saliva creation, decreased hygiene of oral cavity, diabetes decompensation, caries in teeth, sialolithiasis

Symptoms: swelling of gland, erythema and swelling of its duct with discharge of small amount of turbid or suppurating saliva. Skin above gland can be red, painful, in colliquation is present fluctuation with possible spontaneous perforation.

Diagnostics: history, local finding, bacteriology

Therapy: wide spectrum ATB, increased fluids intake, vitamin C. Pilocarpine, increased hygiene of oral cavity

Parotitis recidivans (M.Payen)

Definition: recurrent bacterial inflammation of mostly gl.parotis in children

Symptoms: unilateral painful swelling of gland with decreased production of turbid and suppurating saliva. Recurrences usually stop after adolescence.

Diagnostics: history, clinical course

Therapy: ATB, removal of infectious focus (adenotomy, tonsillectomy, caries in teeth)

Sjógren´s syndrome (myopeithelial sialoadenitis)

Definition: autoimmune disease, afflicts usually women

Symptoms: xerostomia, sclerodermia, panarteritis, keratoconjunctivitis sicca and atrophy of salivary glands

Diagnostics: immunology, biopsy

Therapy: frequent drinking of small amounts of water, saliva production stimulation, corticosteroids according to state

Mikulicz´s syndrome

Definition: autoimmune disease, afflict usually women

Symptoms: atrophy of salivary glands

Diagnostics: immunology, biopsy

Therapy: frequent drinking of small amounts of water, saliva production stimulation

Sialolithiasis

Definition: stone or more stones in duct or salivary gland parenchyma of variable size

Symptoms: swelling and pain of submandibular gland occurs after meal because of obstruction in duct

Diagnostics: history, palpation, USG, X-ray with contrast

Therapy: massage, surgery, ATB therapy in case of inflammation

Complications: abscess in the gland, phlegmona of oral cavity base

Tumors of salivary glands

Benign

- **Ranula** – retention cyst – develops by obliteration of one of the small duct of glandula sublingualis. It looks like painless, mobile formation, usually contains fluid. Can restrict tongue movement or can cause swallowing problems. Ranula can rupture itself.

Therapy: marsupialization or excision

- **Mucocele** – retention cyst of salivary gland in oral cavity mucosa

Therapy: marsupialization or excision

- **Hemangioma** – endothelial proliferation in several months after birth with followed regression in years. More frequent in girls.

Therapy: observation, corticosteroids

- **Vascular malformations** (lymphatic, venous, arterial) – congenital, they grow proportionally with child

Therapy: embolization, excision, exstirpation

- **Pleomorphic adenoma** –possibility of malignant change, recurrences

Therapy: excision, parotidectomy

Malignant

Malignant tumors in comparison with benign cause duct destruction (noticeable in sialography), or paresis of nerves (n.VII). Probability of malignant tumor presence increased with decreased size of salivary gland (parotis – submandibularis – sublingualis – small salivary glands)

- **Rhabdomyosarcoma** – embryonal type in 2-5 years of age, alveolar in adolescents

Therapy: excision, chemotherapy

- **Mucoiepidermoid carcinoma** – if surgical removed it has good prognosis

Therapy: excision, chemotherapy, radiotherapy

Tumors of oral cavity and pharynx

Are rare in children, benign are in 90%. Vascular tumors are present usually in age under 6 years, odontogenous tumors in older.

Examination: history – change of size, pain, bleeding, inflammations, ulcers, speech, swallowing, breathing. Endoscopic examination, CT, MRI in large tumors. Biopsy.

Content

1. Benign tumors

    1.1 Hemangioma

    1.2 Vascular malformations

1.3 Others

2. Malignant tumors

Benign tumors

Hemangioma

Afflicts usually lips, face and tongue. Mucosal hemangioms are red, submucosal blue or violet. Small symptomatic hemangioms have to be observed until involution. Larger can cause pain, bleedin or obstruction of breathing and swallowing – in this time is needed surgery.

Therapy: corticosteroids (30-60% respond), CO2 laser, interferon-alfa-2a

See also Hemangiom in Tumors and expansive processes in children

Vascular malformations

See Vascular malformations in Tumors and expansive processes in children

Others

Mucocele: painless, soft, flat formation on tongue, oral cavity base, in valecula can cause obstruction of airways. Mucocele of oral cavity base is called ranula, can spread on external neck.

Therapy: excision or marsupialization

- **Squamous papilloma**: small, slowly growing formation on palate, tongue or lips

Therapy: excision, recurrences are uncommon with exception of rare oral papillomatosis

- **Dermoid**: usually in ventral part of oral cavity base (20% dermoids of head and neck)

Therapy: excision

- **Choristoma:** histologically normal tissue in uncommon localization (most frequently cyst with gastric mucosa on tongue or oral cavity base)

Therapy: excision

- **Desmoid (fibromatosis**): probably from muscular fascia, locally aggressive, often recurrences

Therapy: wide local excision

- **Lingual thyroid gland**: spherical red formation in middle plane between foramen caecum and valecula. It causes dysphagia, dyspnea, dysphonia and sometimes bleeding. In 70% it is the only functional thyroid gland.

Therapy: autotransplantation

- **Epulis:** congenital tumor from granular cells – hard submucous node

Therapy: excision

- **Neural tumors**: multiple neurinoms of lips and tongue in MEN 2a (together with pheochromocytoma, medullar carcinoma and hyperparathyreoidism). Neurofibroms in patients with neurofibromatosis type I.

Malignant tumors

- **Rhabdomyosarcoma** is usually on tongue, soft palate or face. It expresses itself as a fast growing swelling often with bleeding and ulcers. In the time of diagnostics metastases are usually present in lungs and bones. See also Rhabdomyosarcoma in Tumors and expansive processes in children

- **Other sarcoms:** most frequent firbosarcoma, leiomyosarcoma, angiosarcma, Kaposhi´s sarcoma. Usually develops large infiltrating submucous swellin with bad prognosis

- **Spinocellular carcinoma:** of tongue, lips and palate develops in children on genetic base or in case of immunity defect e.g. after transplantation (no influence of tobacco and alcohol). Children tumors are more aggressive with worse prognosis than in adults.

- **Other tumors:** rare cases of neuroblastoma, melanoma, lymphoma, mucoepidermoid carcinoma, adenoid-cystic carcinoma, hemangiopericytoma

Therapy: radiotherapy 6 500 – 7000 cGy, prophylactic irradiation of neck lymphatic nodes, neck dissection, resection of residual or recurrent tumor. Adjuvant chemotherapy in case of dissemination. 5 years survival in children is 40%.

Injury of oral cavity and pharynx

Content

1. Mechanical injuries
2. Insect bite
3. Acid or alkali burn
4. Foreign bodies in pharynx

Mechanical injuries:

- punctured, cutting, gunshot

Pathogenesis:

- manipulation with sharp items
- fall of child on things in its mouth (pencils, toys, spoon)
- fall on sharp edge of furniture

Diagnostics: history, clinical examination (examination of teeth and jaw is necessary), eventually imaging methods

Therapy: mostly outpatient (ambulatory) – small wounds heal spontaneously, regimen precautions are sufficient (spoon food, chamomile). Larger wounds – suture of wound, according to type of injury administer antibiotics

Insect bite

Wasp or bee bite in this area is dangerous especially for possible great tissue edema or allergic reaction with respiratory problems

Therapy: calcium, antihistaminics, corticosteroids, observation is suitable for possible worsening of patient

Acid or alkali burn

Pathogenesis: drink of lye or acid caused by replacement of bottles, in small children out of curiosity

Symptoms: erythema, tissue coating, ulcers. Sometimes dysphagia, odynophagia or salivation is present.

Diagnostics: history, local state, swallow pathways endoscopy, toxicology center

Therapy: antishock therapy with ATB therapy, by suspicion of swallow etching perform after 24 hours esophagoscopy . Young organism react with global symptoms on local damage more expressively then adult. Development of metabolic breakdown can be faster.

Foreign body in pharynx

Pathogenesis: the most common cause is part of food (fish bones), followed by pins, glass fragments, coins

Symptoms: dysphagia, odynophagia, bleeding, breath difficulties

Diagnostics: history, local state, X-ray

Therapy: foreign body extraction, treatment of wound, possibly ATB. If the foreign body is stuck in hypopharynx or swallow, endoscopical extraction in anaestesia is necessary.

Odontogenous complications in ENT

- **infectious complications**

Odontogenous infections can spread out of gingiva and jaw to the surrounding tissues. Infection can spread from lower jaw to the face, tongue, base of the oral cavity and neck, form upper jaw to the

face and maxillary sinus. In soft tissues can develop abscess or phlegmona. Inflammation can spread even from these parts further to the eye socket, intracranial space or to the mediastinum.

- **non-infectious complications**

After tooth extraction can occur aspiration or swallowing of tooth, surgical instruments or dental plate

Basics of pharyngeal surgery

Adenotomy

is removal of adenoids (pathologically changed pharyngeal tonsils). Now it is made in global anaesthesia, witch leads to decreasing number of complications (bleeding) and repeating operations. Indication for adenotomy is pathology of pharyngeal tonsil (hypertrophy, inflammation)

Tonsillectomy

is total removal of palate tonsils. Most often indications are:

1. recurrent acute tonsillitis
2. chronic tonsillitis (according to patient´s problems)
3. palate tonsils hypertrophy by chronic inflammation or by repeating acute inflammations in history
4. complication of tonsillitis
5. tumor of oral cavity or pharynx (extended tonsillectomy)
6. complete lateral neck fistula form 2. branchial arch (unilateral tonsillectomy

Tonsillotomy

is partial removal of palate tonsils. Indication is obstructive sleep apnea syndrome (OSAS) caused by hypertrophy of palate tonsils. There have to be present no other pathology of tonsils.

Comments to embryology of larynx and trachea

Larynx evolves from cranial end of laryngotracheal tube, which develops from sacciformis evagination on ventral part of ventral gut. Embryonic aditus laryngis has T-shape (at the end of 1.month). Inner part of larynx merge together by entoderm proliferation. Larynx is recanalizating because of cells destruction in central part of tube. Laterally are formed slit shaped basics of ventriculus laryngis. Upper edge if thous slits changes into plicae ventricularesm lower edge into plicae vocales.

Cartilages and muscles of larynx are evolving from 4.and 5.branchial arch. Muscles are innervated by n.X.

Larynx is formed cranially and later descends caudally. In 5.fetal month is in the level of nasopharynx. Neonate has epiglottis in the level of 2.-3.neck spondyl, adult in 5.neck spondyl. Growth is faster during pubescence, especially in boys.

Trachea evolves from laryngotracheal tube after larynx. Cartilages are differentiating in 2.month. Larnygotracheal tube has dead end with button-like bronchopulmonal entodermal evagination. Lungs and bronchi are formed from it.

Clinical anatomy of larynx and trachea

Content

Clinical anatomy of larynx

Neonates have small and wide larynx. Average diameter in adults is 5 cm in women and 7 cm in men. This difference causes different voice in women and men. Vocal cords lenghten by 3 mm in girls and 5-10 mm in boys during pubescence. Lenghtening of vocal cords causes mutation voice disorders. Larynx has final size in 23 years of age. Voice aging is present around 60.year of age (calcification of cartilages), vocie range and volume is decreased, voice color is more sharp.

Larynx is located on ventral part of neck and its ventral part is covered by caudal muscles of hyoid bone (infrahyoid muscles) and by both leafs of infrahyoid fascia. On lateral walls are lobes of thyroid gland. Behind larynx is hypopharynx. Ventrocranially is hyoid bone and tongue. Caudally continues as a trachea. Laryngeal wall consists of cartilages connected by fibrous tissue and joints. On their frontal, lateral and dorsal walls are muscles which move the cartilages and controll tension and distance of vocal ligaments. Larynx is covered with ciliary epithelium, its cilia oscillate towards entrance. On vocal ligaments and epiglottis is spinocellular epithelium. In mucosa are small glands. Lymphatic tissue is abundant, especially on ventral part of epiglottis and in ventriculus laryngis. Submucous fibrous tissues are thin, so that infection can cause edema and resulting dangerous striction of larynx interior.

Laryngeal skeleton

Contains 3 unpaired cartilages and 2 pair.

- **cartilago thyreoidea** – has right and left leaf
- **cartilago cricoidea** – supporting laryngeal cartilage.
- **Epiglottis**
- **Cartilagines arytenoides** – are connected by joint with upper part of annular cartilage. Ligamentum and muscle of vocal cord are bound to processus vocalis (ventral processus). Arytenoid cartilage is moving and couses dilatation and closure of vocal cords fissura, which is called glottis

Cavum laryngis

- **supraglottis:** contains all anatomic structures above vocal cords (laryngeal part of epiglottis, aryepiglotic plicae, ventricular plicae, sinus Morgagni, arytenoid area)
- **glottis:** vocal cords
- **subglottis**: area under vocal cords, continuing to trachea

Muscles of larynx

Are striated muscles, richly innervated and localized around larynx in 3 groups:

Ventral:

- **M.cricothyreoideus** draws cartilago thyreoidea to annular cartilage. Causes tension of ligamenta vocalia.

Dorsal:

- **M.cricoarytenoideus dorsalis** (posticus) – draws processus muscularis cartilaginis arytenoidei medially and causes tension of vocal ligaments – abductor of vocal ligaments
- **M.arytenoideus** (transversus) – adductor of vocal ligaments
- **M.aryepigloticus** – bends epiglottis forward

Lateral: sphincters of glottis

- **M.cricoarytenoideus lateralis** – antagonist of similar dorsal muscle:
- **M.cricoarytenoideus dorsalis**
- **M.thyreoarytenoideus** has external part (pars lateralis) and inner part (pars vocalis - m.vocalis). It is localized in plica vocalis. It is antagonist of m.cricothyreoideus

- **M.thyreoepiglotticus** – same function as m.aryepigloticus

Vascularization of larynx

- a.laryngea superior from a.thyreoidea superior
- a.laryngea inferior from a.thyreoidea inferior
- veins are called similarly
- lymphatic vessels drain lymph deep neck lymphatic nodes.

Innervation of larynx

- **motoric**: n.laryngeus inferior (n.recurrens) from n.X. On the right side goes under a.subclavia dx. and goes up between trachea and esophagus to the larynx. On the left side is this nerve lower, goes under aortal arch and goes alongside trachea to larynx. Righ one is shorter than left one. The only exception is m.cricothyreoideus, it is innerved from n.laryngeus superior (branch of n.X as well)
- **sensitive:** n.larnygeus superior

Physiology of larnyx

- **Respiratory function:** larynx maintain flow of passing air. During breathing is glottis free and has triangle shape. During ispiration is little wider, during exspiration little narrower. Velocity of air is 3-5 m/sec, during scream 30-40 m/sec.
- **Phonatory function:** human voice is formed by periodic oscillation of air column above vocal cords. Primary tone is formed by opening and closing the glottis due to air pressure changes. Speech cavities are: nasal cavity (its shape doesn´t change), pharyngeal cavity (its shape does change and play role in speech sounds formation). Main role in speech sound formation play oral cavity, its shape can be easily changed. Voice and speech is communication controled by hearing, thats why hearing disorders are followed by speech disorders.
- **Protective function:** maintained by muscles and laryngeal mucosa. Kymphatic tissue in larnyx has similar function as in pharynx. Glands produce mucus and ciliary transport is responsible for transport of mucus and small foreign bodies towards oral cavity. Protective reflexes (coug and swallowing) are controled by n.X. Cough reflex is caused by irritation of sesitizve parto of n.X in larynx, pharynx and trachea. It helps to clean not only the larynx itself, but whole tracheobronchial tree as well and helps to remove foreign body.

  During swallowing larynx protects lower airways. Prevents intrusion of food to the lower airways by coordinated movements of muscles of larynx, oral cavity an tongue. During swallowing larynx is moving proximally and radix linguae dorsally.

Clinical anatomy of tracheobronchial tree

Trachea is a tube connected cranially with larynx. Ligamntum cricotracheale binds it to annular cartilage. Trachea begins in the level of C6 and ends in the level of Th 4-5 as a bifurcatio tracheale –

bronchus dexter and sinister. Carina tracheae is sagital horizontal edge protruding in the bifurcation into tracheal interior. In the walls of trachea are approx. 15 hyallinous cartilages – cartilagines tracheales.They look like horseshoe and are open dorsally. Dorsal part – pars membranaceus is formed by collagenous and elastic fibrils and smooth muscles. Cartilages ar connected together by ligamenta annularia

Mucosa is pink wit longitudinal plicae in paries membranaceus. It is covered by cilliary epithelium. In epithelium are many calyciform cells. Submucous tissue is thin and contains many glandualae tracheales. Adventitia connects trachea with esophagus and bind it to the surrounding structures.

Bronchi is name for branched system of tubes leading air form trachea into lungs

- **Bronchi principales** (main) are two: left and right. They begin in the bifurcation tracheae. Right bronchus is shorter, wider and more straight than left one.
- **Bronchi lobares**: lobe bronchi (superior, medius, inferior dexter and superio, inferior sinister)
- **Bronchi segmentales**: branches of lobar bronchi
- **Bronchioli:** branches of smallest bronchi, bronchioli terminales

Vascularization and innervation

- Arteries: rr.tracheales from a.thyreoidea inferior and rr.bronchiales from thoracic aorta
- Veins of trachea bring blood into esophageal veins, v.thyreoideae inferiores and vv.brachicephalicae
- Lymphatic vessels leads to nodi tracheales and tracheobronchiales superiores, dexter and sinister, after that to the truncus bronchomediastinalis dexter and sinister
- Nerves are from n.X and neck sympaticus

Physiology of tracheobronchial tree

Air passes through upper and lower airways. Pulmonary artery goes alongside bronchial tree, so the smallest arteries goes together with terminal bronchioli. Arteriols accompany bronchioli respiratorii and capillaries spins around alveols in form of nets. Capillaries are in interstitium of alveolar septum and are tightly connectedy with lamina basalis of alveolar epithelium. Oxgen goes from alveolar air into capillaries and CO2 goes from capillaries into alveols. On the inner parts are located alveolar macrophages – dust cells, which contains phagocytated dust particles and toghether with macrophages inside interstitium form defense against infection. Secretory pneumocytes form surfactant, which decreases surface tension and prevents collapse of alveols – pulmonary atelectasis.

Examination methods of larynx and trachea

Aspection, palpation

With sight we examine ventral part of neck and with palpation during swallowing, speaking and in rest.

Laryngoscopy

- **indirect**: by larnygoscopic mirror or by magnifying optical laryngoscope. Picture is vertically reversed.

- **Direct**: by rigid tube and microscope (for endoscopic surgery) or by flexible laryngoscope. Picture is horizontally reversed.

Laryngoscopy serves for examination of anatomic changes in larynx and function of laryngeal interior. Patient must say eee or iii during eamination. Special case is examination of correct phonation with help of stroboscopic light.

Examination of tracheobronchial tree

- **history:** dominant symptoms are dyspnea, cough, hoarseness, stridor, swallowing problems, odynophagia. We must find out time of beginning, frequency and intensity.

- **Physical examination:** auscultation, functional pulmonary tests (tidal volume, residual volume, Total lung capacity (TLC), Forced vital capacity, Functional residual capacity, Forced expiratory volume at 1 second, The ratio of forced expiratory volume at 1 second to forced vital capacity), polysomnografie (EMG, EEG,EKG, electrooculografie, nasal and oral airflow, re-spiratory movements and effort, saturation, position during sleeping.

- **Imaging methods:**

- **X-ray:** ventral and lateral picture is necessary for screening in suspicion on airways disease. Lateral picture can be made in exspirium or inspirium. X-ray is suitable for diagnostics of pathology in retropharyngeal space (infection, abscess, tumor). Subglotic or tracheal stenosis can be ssen on X-ray. Epiglottic edema can be seen on lateral picture. Normal picture is suitable for identification of foreign body. We can see contrast bodies, non-contrast can cause obstruction or stricture in airways.

- **Fluoroscopy:** it is used for diagnostics of sleep apnea syndrome

- **Contrast radiography:** X-ray with help of contrast fluid. It is used for diagnostics of GERD, aspiration, tracheoesophageal fistula or foreign body.

- **Arteriography:** used for detection of great vessels anomalies or vascular tumors

- **CT:** in axial and coronary projection. IT is method of chice in case of injuries, congenital abnormalitites, tumors and foreign bodies. CT with contrast helps us to find vascular lesions, abscesses, etc. Disadvantages are higer price, irradiation, static picture. Ultrafast CT allows dynamic examination of airways

- **Virtual bronchoscopy:** is used in severly ill patients, in case fo coagulopathy, for diagnostics of tumors, chronic foreign body, etc. It is non-invasive method without need of general anaesthesia. It is 3D reconstruction of CT scans (1mm thickness). Examination can be made with contrast fluid (tumors, extraluminal pathology). For diagnostics of tracheomalatia or airways collapse can be used dynamic inspiratory-exspiratory imaging.

- **MRI:** has same advantages as a CT. It doesn´t irradiate the patient and can display anatomic structures in many different levels, has excellent resolution. IT is not suitable for bones examination, CT is much better.

- **Ultrasonography (USG):** suitable for examination of solid formations, cysts, abscesses, lymphadenopaties, which can cause strictures of airways. In last few years we have high frequency endoluminal USG

- **Nuclear methods**: especially for diagnostics of tumors (SPECT, PET)

- Bronch**oscopy:** endoscopic examination of trachea and bronchial tree. We can divide bronchoscopy into rigid and flexible. Bronchoscopy can be diagnostic or therapeutic.

- **Rigid bronchoscopy**: is made in general anaesthesia. Rigid tube is inserted through mouth, patient is laying on the back. Orientation points are uvula and epiglottis. We can see glottis and after insertion of tube between vocal cords we can see trachea and branching of bronchial tree. Patient is intubated by bronchoscope and simultaneously is made the operation (foreign body removal, stenosis dilatation, partial disection of tumor). Foreign bodies are removed only by rigid bronchoscopy in general anaesthesia.

- **Flexible bronchoscopy**: used for dignostics of tracheobronchial tree diseases. In children are made in general anaesthesia. Tube can be inserted via mouth or nose. Orientation points are same as in rigid bronchoscopy. If we found any pathology, we can do diagnostic procedures – biopsy, irrigation, aspiration, abrasion, etc.)

Congenital defects of larynx and trachea

Early diagnostics of congenital defects is condition of early curative surgery and even life salvation. Congenital defects are diagnosed according to fonační and respiratory dysfunction. Essential are laryngoscopy, X-ray, CT and pediatric examination.

Stridor laryngis congenitus

Definition: temporary stridor in children, caused by laryngomalacia

Etiology: congenital defect, caused by hypoplastic ligaments of sucklings larynx and epiglottic cartilage (laryngomalacia).

Pathogenesis: during inspirium are supraglotic structures sucked in the laryngeal interior and distinct inspiratory stridor is present, expirium is silent, voice is plain. The epiglottis is tipped over larynx entrance by the flow of expired air. By this mechanism the laryngeal interior is narrow and air which goes through this slot causes inspiratory stridor

Symptoms: inspiratory stridor, during crying or cough is stronger, by the change of position can be reduced. The strongest symptoms are immediately after birth, after that will subsequently and spontaneously diminish.

Diagnostics: history, laryngoscopy

Therapy: symptomatic (patient positioning, air moistening, AD vitamins). Stridor spontaneously vanishes around 18th month of child age, ligament structure of larynx gets stronger.

Complications: retraction of jugulum and intercostal muscles, cyanosis during crying. Suffocation and phonatory problems aren´t present.

Differential diagnostics: all other congenital defects

Laryngocele

Definition: laryngeal mucosa evagination full of air and mucus. It is localized in meatusriculus larnygis. We differentiate inner and outer one.

Etiology: rare congenital defect

Pathogenesis: outer one evaginates outwards through membrana thyreohyoidea and is palpable on lateral part of neck. It looks like cyst, but it increases during Valsalv´s maneuver. Inner one develops as a hernia of meatusriculus laryngis.

Symptoms: are same in both types: hoarseness, dyphagia, sometimes dyspnea. In case of forced expirium we can see bulge on lateral part of neck.

Diagnostics: laryngoscopy, X-ray or CT (we can see air or contrast fluid inside)

Therapy: surgical – resection.

Complications: breathing and swallowing problems

Differential diagnostics: cyst doesn´t change its shape

Diafragma laryngis

Definition: membrane in glottis area, it causes severe dyspnea

Etiology: stopped larynx evolvement in 10.embryonic week

Pathogenesis: fibrous membrane is localized in frontal 1/3 between vocal cords. Mambrane can be in supra or subglottic area and has variable thickness

Symptoms: aphonia, breathing problems and stridor. They depends on size of membrane.

Diagnostics: history, symptoms, laryngoscopy

Therapy: tracheostomy is recommended, because discisions are healed by scars, which causes strictures of larynx. Definitive solution is made in older age.

Complications: respiratory infection, swelling of airways, respiratory failure

Differential diagnostics: other congenital defects of larynx

Inflammations of larynx and trachea

Content

Laryngitis acuta

Laryngitis catarrhalis acuta

Definition: acute inflammation of laryngeal mucosa only

Etiology: viral origin

Pathogenesis: disease starts with rhinosinusitis, flu, as a descendent catarrhus of upper respiratory pathways

Symptoms: iritating cough, light dysphonia, light inspiratory stridor, drought and burning in neck

Diagnostics: history (viral infection), indirect laryngoscopy (vascular laryngeal mucosa, edematous vocal cords with mucus)

Therapy: therapy of basic viral infection, antipyretics, fluids, vitamin C, antihistaminics, spare vocal cords. If bacterial superinfection is present antibiotics are necessary

Complications: commonly night dyspnea with vocal cords spasm, bacterial superinfection and development of acute bacterial laryngitis

Laryngitis acuta subglottica

Definition: severe and dangerous form of acute laryngitis which affects submucous fibrous tissue in subglotic space of larynx

Etiology: viral (influenza, parainfluenza) vith possible bacterial infection

Pathogenesis: inflammation with submucous infiltration. There is sparse submucous tissue in subglotic space, which can easily tumify. Developed swelling causes larynx diameter striction.

Symptoms:

- Always present: dyspnoe, inspiratory stridor, dysphonia, barking cough
- Sometimes present: dysphonia, cough

Diagnostics: history, laryngoscopy (edematous, red mucosa in form of subglotic stenosis)

Therapy:

- first aid: cold wet air, cold-wet compress, patient must sit, lukewarm tea
- medication: corticosteroids (Rectodelt supp.), adrenaline inhalation, antihistaminics, antitustics, expextorantia. Antibiotics in case of bacterial superinfection
- secure airways: intubation or tracheostomy

Complications: inflammation progression with daily dyspnea, vocal cords inflammation, laryngotracheobronchitits

Epiglottitis acuta

Definition: severe inflamation on submusous tissue, localized behind epiglottis

Etiology: Haemophillus influenzae, group B

Pathogenesis: Children infectious diseases or larynx injury leads to flegmona. In children it looks like serous mucosa inflamation with difuse swelling. It is substantially more rare than laryngitis subglotica. Flegmona is commonly localized on epiglottis and can cause abscess.

Symptoms:

- always present: dyspnoe, inspiratory stridor, larynx pain, retraction of auxiliary breathing muscles (jugulum, intercostal, epigastrium), cyanosis, paleness, fever, dysphagia, odynophagia, salivation
- sometimes present: dysphonia, cough

Diagnostics: history, symptoms, laryngoscopy (edematous red epiglottis)

Therapy:

- first aid: lay on guts, koniotomy
- secure airways: intubation, koniotomy or tracheostomy
- medication: antibiotics, antiedematous therapy (corticosteroids), vital functions monitoring

Complications: epiglotical abscess. In this case yellowish cap can be seen on the lingual part of epiglottis. Incision and dilatation is necessary. There can be even the unconfined form with septic state, inflamation can spread to the mediastinum or cartilage and cause perichondritis.

Prevention: vaccination (obligatory in Czech republic)

Laryngitis oedematosa

Definition: inflamation of laryngeal mucosa with serous infiltration of submucous tissue

Etiology: laryngeal swelling can develop during larynx or perilaryngeal inflamation (paratonsillary or parapharyngeal abscess). Non-infectious swelling can develop after injury, due to burning, foreign body, by children due to allergy or gastrooesophageal reflux disease (GERD)

Pathogenesis: transsudate caused by lympho- or venostasis infiltrates laryngeal fibrous tissue in places, where it is very thin (epiglottis, subglottis, aryepiglotical plicae)

Symptoms: acute dyspnoe, inspiratory stridor, suffocation

Diagnostics: history, symptoms, laryngoscopy (pale swelling)

Therapy: hospitalization, ATB, corticosteroids, antihistaminics, calcium, intubation or tracheostomy

Laryngotracheobronchitits pseudomembranacea et crustosa

Definition: inflamation of larynx, trachea and bronchi with mucosal swelling and pseudomembranes

Etiology: bacterial superinfection (Staphylococci) after viral inflammation (flu, measles)

Pathogenesis: airways obstruction is caused by mucosal swelling, exsudate (which gets dry and forms crusts) and pseudomembranes (contains fibrin, leucocytes, epithelies, microbs) In case of severe form inflamation is spreading from larynx to trachea and bronchi. Total obstruction with atelectasis or valve occlusion with emphysema can develop in bronchi. Airways obstruction or sepsis can lead to suffocation or even death of child.

Symptoms: child has breathing problems, is suffocating inspiratory or expiratory stridor is present. Child retracts epigastrium, jugulum, supraclavicular area, intercostal muscles or possibly whole chest during insipirum. Child is pale or cyanotic, restless, has fever and can have dysphonia.

Diagnostics: history, laryngoscopy (red, edematous mucosa with thick mucus and sometimes with pseudomembranes and crusts)

Therapy: hospitlization is necessary, ATB, corticosteoids, antihistaminics, cardiotonics, calcium, oxygen, moisten air. In case of worsening intubation or tracheostomy is required. According to state of children bronchoscopy with mucus and crusts removal can be needed.

Laryngitis chronica

Definition: chronic inflammation of laryngeal mucosa, can be hypertrophic or atrophic. Practically not present in children.

Etiology: repeated nasal mucosa, pharynx or paranasal sinuses inflammation, allergy, frequent laryngitis, flegmonous mucosal inflamation, GERD, surgery of larynx, hyperthyreosis, diabetes mellitus, anemia, chronic kidney diseases, hormonal changes, environmental factors, result of burning

Pathogenesis: fibrous tissue hyperplasia in laryngeal mucosa, slime glands and lymphatic tissue atrophy

Symptoms: hyperplastic mucosa can cause light respiratory or voice problems. The mucus gets dry on vocal cords during atrophic laryngitis and cause hoarseness and cough.

Diagnostics: history, laryngoscopy

Therapy: cause of chronic laryngitis has to be found out. This cause has to be eliminated, spa therapy and airways moistening is recommended

Complications: this disease is considered as a precancerosis

Specific laryngeal inflammation

Etiology: mycobacterium tuberculosis (TBC), treponema pallidum syphillis), klebsiella ozenae or rhinoscleromatosis

Pathogenesis: systemic disease witch can affect laryngela mucosa

Symptoms:

- TBC has milliary and infiltrative form with nodes and ulcers on mucosa. In chronic infiltrative form are vocal cords edematous and red.
- Syphillis - larynx has ulcers, infiltrates and enanthema
- Other specific inflammations are very rare

Diagnostics: serology, history, laryngoscopy

Therapy: therapy of given disease

Complications: depends on the disease

Tumors of larynx and trachea

Content:

1. Benign
    - 1.1 Hemangioma
    - 1.2 Polyp
    - 1.3 Papiloma
    - 1.4 Other tumors

2. Malignant

Benign tumors of larynx and trachea

Hemangioma

Pathogenesis: tumor can be found on different places, most often is found in subglotic area on the side. Size varies, tumor can spread to pharnyx or cervical spondyls. Hemangioma is more often in girls.

Symptoms: subglotic hemangioma causes stridor, sometimes dysphonia and dyspnoe

Diagnostics: history, CT, angiography, laryngoscopy (blue-red formation). Biopsy cannot be made because of bleeding risk.

Therapy: embolization, corticosteroids. Hemangioms spontanneously involve in the age of 2-3 years. Tracheotomy if airways obstruction is present. Radiotherapy is not recommended, because pf imminent larynx growth disorder and risk of thyroid gland carcinoma development. Endoscopic cryotherapy can cause striction. Surgical ablation via laryngeal fissure is possible, as well as laser ablation.

Complications: dysphonia, dyspnoe, dysphagia

Polyp

Definition: organized haematoma

Etiology: smoking, voice overstrain

Pathogenesis: polyp develops due to chronic irritation of mucosa after voice overstrain, it is impaired by smoking or environmental factors. The result is haematoma organization.

Symptoms: dysphonia, incerased voice fatigue

Diagnostics: laryngoscopy (widely attached formation in fronat and middle third of vocal cord, it has smooth surface and pale or red colour), history

Therapy: voice using practice, microlaryngoscopic ablation

Papillomatosis

Definition: it is the most common benign tumor of larynx in children

Etiology: human papilloma virus (HPV-6, HPV-11)

Pathogenesis: virus is transmitted due to bad hygiene or from mother to child during birth. There are about 50 types of virus. HPV causes warts, papiloms in oral cavity and nose or condyloms in anogenital area

Symptoms: hoarseness, dysphonia, aphonia, inspiratory stridor, dyspnoe, skin papiloms or condyloms in anogenital area are often present.

Diagnostics: history, laryngoscopy (pink or pale formation commonly localized on vocal cords, can be found in supra- or subglotic part of larynx as well). If a child has a persistent dysphonia or repeating laryngitis, laryngoscopy in anaestehis is indicated.

Therapy: microlaryngoscopic o surgical ablation. If patient receives conservative treatment (ATB, hormons, vitamins, cytostatics, interpheron), reccurence is frequent.

Other tumors

Fibroma, lipoma, myxoma, neurofibroma, …

Symptoms: dysphonia and stridor – depends on size and localization of tumor

Diagnostics: histological examination

Therapy: microscopic surgery, voice training after ablation is necessary

Malignant tumors of larynx and hypopharynx

Definition: most often are carcinoma and sarcoma

Etiology: smoking (smokers especially with more than 20 cigarettes per day for 20 years, 90% are men), alcoholism. Malignant tumors are exceptional in children.

Pathogenesis: Most often is spinocellular carcinoma. TNM classification describes disease stage and is only for carcinoms:

T category: extent of primary tumor

N category: presence of tumor in regional cervical lymphatic nodes

M category: presence of distant metastases

From these categories we can assess disease stage on early stages (I, II) and late stages (III, IV)

Symptoms: depends on size and localization of tumor. Most frequent is hoarseness. Earliest presence of hoarseness is in case of tumor localized on vocal cords. About 50% of malignant tumors are localized in glotic area. Because of early hoarseness it can be earlier diagnosed (and cured) and that´s why it has better prognosis. Metastases are rare. Supraglotic carcinoma is present in spirits drinkers and smokers. Tumors of laryngeal entrance have odynophagia as a first sign. Every patient with hoarseness or swallow problems longer than 3 weeks should have laryngoscopy, because those symptomes are often disregarded. In late stage cough, haemoptysis, dyspnoe or suffocation are present.

Diagnostics: history, microlaryngoscopy and histological examination. Diagnosis must be histologically confirmed.

Therapy: Disease stage and histopatological grading is important for therapy and prognosis. Grading is level of tumor cells. IT is described from G1 to G4. Therapy is surgical and oncological (radiotherapy and chemotherapy) or combination of both.

Surgical therapy: larnygectomy

Prognosis: early stages of glotic form of carcinoma has good prognosis. Supraglotic and subglotic forms come usually in later stages so the prognosis is worse (in addition those types have early metastases).

Injuries of larynx and trachea

Content

1. Injuries of larynx
    - 1.1 External injuries of larynx
        - 1.1.1 Blunt injury
        - 1.1.2 Sharp injury
    - 1.2 Inner injuries of larynx
        - 1.2.1 Burning (acid or alkali) and scalding
        - 1.2.2 Aspiration
        - 1.2.3 Stenosis caused by intubation
        - 1.2.4 Injury of cricoarytenoid articulation

Injuries of larynx

External injuries of larynx

Blunt injury

Strike with blunt weapon on ventral part of neck.

- commotion – edema of mucosa

symptoms: dyspnea, cough

Therapy: corticosteroids

- contusion – damage of submucous tissue

symptoms: dyspnea, cough, hematoma

therapy: corticosteroids, securing airways

- compression – fractures of cartilages

symptoms: dyspnea, cough, emphysema, hematoma

therapy: corticosteroids, ATB, securing airways

Sharp injury

Gunhot, stab or cutting

Therapy: surgery, ATB, corticosteroids

Inner injuries of larynx

Burning (acid or alkali) and scalding

Aspiration of corrosive substances, gases, hot steem

Symptoms: pain during speech and swallowing or even in rest, subcutanneous emphysema, bleeding, dyspnea

Therapy: securing airways, surgery, corticosteroids, ATB

Aspiration

Definition: aspiration of foreign body form oral cavity through larynx into trachea and bronchi. We divide them into acute and chronic.

Etiopathogenesis:

- undeveloped teeth, infection of airways, unrest during eating (usually peanut, vegetable or toys)
- games, insufficient number of upper limbs (pin, nails, ...)

Majority of foreign bodies (60%) are localized in right bronchus (it is larger and more straight than left one)

Symptoms:

- dyspnea, irritating cough, cyanosis, suffocation
- minimal symptomatology (during this phase patient attends doctor!!)
- bronchopneumonia

According to foreign body position can afflicted lung ventilate physiologically or partial - valve closure can develop. In this case inhaled air passes into lung but cannot go out and this results in pulmonary emphysema. In case of total closure develops atelectasis. IF foreign body stays long time in bronchus, it causes inflammation and granulation tissue formation. There are recurrent suppurating bronchitis, pneumonia, atelectasis or even pulmonary abscess. These symptoms are more significant and often in organic foreign bodies.

Diagnostics:

- history – needed for bronchoscopy indication
- clinical ENT examination, lungs auscultation
- imaging methods: X-ray – can reveal foreign bodies. We can see atelectasis or emphysema in non-contrast bodies
- bronchoscopy

Therapy:

- first aid: Heimlich´s maneuver (cannot be used in pregnant women and young children), koniotomy (foreign bodies locaed near vocal cords)
- bronchoscopy with foreign body extracion, ATB in case of inflammation
- thoracotomy (if bronchoscopic extraciton is unsuccesfull)

Stenosis after intubation

Specific group of diseases forms long lasting intubation. Endotracheal or tracheostomic cannula cause tracheomalatia (caused by cannula´s ballon pressure on the tracheal wall) with cicatrication and stenosis or even fistula development. Postintubation granulomas are formed in dorsal part of glottis.

Therapy: ballon dilatation, trachemotomy, tracheoplasty

Injury of cricoarytenoid articulation

Etiology: it is usually caused by injury of cricoarytenoid articulation during intubation. Intubation can cause luxation of this articulation. This articulation can be afflicted in case of stenosis in dorsal commisura as well.

Symptoms: restriction of voice efficiency, persistent hoarseness, dyspnea in case of stenosis, however there can be no symptoms at all.

Diganostics: during laryngoscopy we can see vocal cord in medial position, arytenoid protuberance in larynx interior (similar picture as in unilateral affliction of n. Laryngeus recurrens), peroperative examination of arytenoid cartilage mobility, EMG of larynx with normal findings (without signs of denervation or reinervation).

Therapy: mobilization or reposition of arytenoid cartilage, often unsuccesfull becaus of fibrotization in the vicinity of articulation

Differential diagnostics:

- unilateral paralysis of vocal cord – EMG with signs of denervation or reinervation, arytenoid cartilage is normally mobile
- rheumatoid arthritis – in history we miss tracheal intubation. Rheumatologic examination is needed.

Larynx inervation defects

Unilateral vocal cord paralysis

Etiology: the most frequent cause is iatrogenous damage of n.laryngeus recurrens during thyroid gland surgery. Another possibility is invasion of n.vagus or n. laryngeus recurrens by tumor (of cranial base, thyroid gland, oesophagus, lungs) or pressure of enlarged lymphatic nodes (lymphoms and metastases of malignant tumors). If previous causes are excluded, unilateral paralysis is described as a idiopatic (common cause is viral or another inflammatory damage)

Symptoms: bilateral paralysis: usually sudden aphonia or dysphonia. Typical is great dyspnoe and voice disability. Dysphagia of fluids can be present. If damage of n.vagus is localized before n.larnygeus superior, risk of aspiration is increased because of laryngeal anaesthesia. Dyspnoe in unilateral paralysis is rare (it is present only in patients with need of higher positive endrespiratory pressure – e.g. astmatics). Patients have rather feel of breathlessness during speech because of insufficient glottis closure.

Diagnostics: voice examination and/or voice analysis. Vocal cord immobility is seen during laryngoscopy. Vocal cord is in paramedial position, arytenoid tuber of afflicted side is sucken into larynx interior. Absence of tumor must be confirmed, if trauma or iatrogenous damage is excluded. Ventral and lateral X-ray chest is a must. In case of uncertainity CR or MRI has to be made. Possible is electromyography (EMG) of laryngeal muscles (m.cricothyreoideus and m. thyreoarytenoideus) by percutanneous needle. EMG is used for diagnostic and prognostic reasons.

Therapy: If the cause is idiopathic or sometimes iatrogenous, paralysis can be transitory and fast spontanneous recovery follows. If the paralysis is long lasting or permanent, therapy depends on patients demands on voice quality. To make voice more sonant glotis closing has to be made by compensatory hyperabduction of afflicted vocal cord to the good one. Sometimes it happens spontaneously, sometimes voice training is necessary. Surgery is indicated where voice is unsatisfactory even after training and there is no chance to movement recovery (after more then 1 year, or confirmed by EMG). It depends on age of pacient, because atrophy of m.vocalis will develop after several years from start and glotis insuficience will be worse and cannot be compensated.

Differential diagnostics: injury of cricothyreoid joint must be excluded. Vital information will offer EMG.

Diferential diagnostic of dyspnoe

Definition: dyspnoe is difficult breathing caused by i.a. airways constriction, folowed by stridor (murmur developing when air passes through constricted airways)

Classification:

- Inspiratory stridor: caused by constriction of larynx and trachea (to the bifurcation). Dysphonia is often present.
- Expiratory stridor: caused by constriction of trachea and bronchi (behind bifurcation). Voice is clear.
- Inspiratory and expiratory stridor: airways obstruction spreads from larynx to bronchi or vice versa (laryngotracheobronchitis)
- Pharyngeal stridor: caused by nose or pharynx obstruction (mucus swelling, adenoids, polyps, tonsils hypertrophy)

Type of stridor helps to find approximate localization of obstruction. Voice volume and intensity can change, e.g. during work. Stridor can develop slowly or appear suddenly. If oxygenation is insufficient, breath gets deeper and slower and auxiliary muscles start working. Sucklings have alar breathing (simultanoeus nostrils movement with breath). Cyanosis, exhaustion, suffocation, unconsiousness and arrhytmia appear after some time.

Differential diagnostic in children:

- Amniotic fluid aspiration
- Asphyctic syndrome (defective intrauterine breathing transformation to postnatal form).
- Congenital defects of airways (choanal atresia, congeital tumors of nasopharynx, laryngeal or paralaryngeal cysts, atresia or stricture of larynx and trachea, lung hypotrophy, etc.), swallow pathways (macroglossia, ductus thyreoglossus cysts, etc.), heart or large veins anomalies, congenital mediastinal tumors, diaphragmatic hernia, congenital defects of central nervous system (hydrocephalus, bulbar paralysis), perinatal damage of CNS (bleeding)
- Aspirated foreign bodies
- Laryngeal inflammation (laryngitis, epiglotitis, laryngeal phlegmona laryngeal abscess, etc.)
- Injury of airways
- Paralysis of laryngeal muscles (central damage, n.vagus branch damage, myopathia)
- Tumors of larynx and lower airways
- Diseases of pharynx and oral cavity (abscess, phlegmona, tumor, OSAS – obstruction sleep apnea syndrome, etc.)

Therapy: secure airways, resuscitation, intubation, bronchoscopy, tracheostomy

Basics of laryngeal and tracheal surgery

Content

1. Microsurgery of larynx
2. Laryngectomy

3. Coniotomy, coniopunction
4. Tracheotomy

Microsurgery of larynx

Definition: removing or treatment of lesser pathology during direct laryngoscopy with help of operational microscope. Pay attention to save function. Possible is use of laser, kryotherapy or local chemotherapy.

Laryngectomy

- Partial laryngectomy horizontal: epiglotectomy
- Partial laryngectomy fronto-lateral: is made in vertical level if glottis is afflicted. Partial laryngectomy can be made by external approach or by endoscope during direct laryngoscopy. Chordectomy is the most frequent vertical partial laryngectomy.
- Total laryngectomy: during this whole larynx is removed (extensive tumor affliction in III. or IV. stage) and tracheostomy is made. Regional lymphatic nodes have to be always checked. If they are histologically positive, neck dissection have to be made. By total laryngectomy are disrupted respiratory function of larynx and voice, so we must substitute them. Respiratory function is substituted with permanent tracheostomy, voice can be substituted with 3 possibilities: (esophageal voice training, electrolarynx or implantation of voice protesis)

Coniotomy, conipunction

Definiton: cut throught or thrust through ligamentum conicum (thyreocricoideum)

Indications: first aid in apneusis caused by pathology localizated cranially from ligamentum conicum

Tracheostomy

Definition:

- Tracheotomy: cut through trachea usually from horizontal or possibly vertical incision. We must spare first and second tracheal ring because of frequent development of subglotic striction
- Tracheostomy: trachea opening in the neck via tracheostomic cannula
- Punctured tracheostomy: this usually do anestesiologist under control of flexible optic. It is kontraindicated in small children due to small size, mobility and flexibility of trachea

Classification:

- upper tracheotomy: above thyroid isthmus
- middle tracheostomy: after cut through thyroid isthmus
- lower tracheostomy: under thyroid isthmus

Indications:

- long lasting intubation
- airways dead space reduction
- airways obstruction
- total laryngectomy

Comments to embryology of esophagus

Esophagus is evoloving form part of ventral gut caudally from pharynx after separation from laryngotracheal tube. Embryonic esophagus is short at first and caudally continues as a base of stomach. In 2.month grows faster, lenghtens and drags entoderm into its interior. Esophagus rotates because of stomach rotation (asymetric course of n.X). Epithelium is forming from entoderm, it proliferates and can fill all the interior of esophagus. Later is forming star-shaped lumen. Fibrous tissue and muscles are forming from mesenchym. Striated muscles in upper third of esophagus is forming from mesenchyme of brancial arches, smooth muscles in lower 2/3 of esophagus are forming from splanchnopleural mesenchym.

Clinical anatomy of the esophagus

Content

Clinical anatomy of esophagus

Esophageal wall has 4 layers: mucosa, submucous tissue, muscles and adventitia

Mucosa forms longitudinal plicae and is formed by spinocellular epithelium

Muscles have 2 layers: inner circular and external longitudinal. Above upper sphincter (Kilian´s) forms m.constricotr pharyngis weak spot, where can develop Zenker´s diverticulum.

Esophageal wall is 2-5 mm thick. Esophagus is 25-30 cm long in adult. Its beginning is in the level of annular cartilage and C6. Esophagus goes in front of spine. It enters stomach in the level of Th 11. Kilian´s sphincter and carida have their own closing mechanisms with constant tonus, which prevents intrusion of air into stomach and reflux of gastric fluids.

Esopahgeal strictions:

- **upper** – pars fundiformis of pharyngeal sphincter and Kilian´s sphincter. Distance from lower incisive teeth is 7 cm in suckling, 12 cm in 10 years old child and 16 cm in adult

- **middle** – caused by pressure of aortal arch an left main bronchus
- **lower –** in the end of esophagus and it is caused by cardia and diaphragm crossing. Distance from incisive teeth is 21 cm in sucklings, 27 cm in 10 years old child and 40 cm in adult

Topography of esophagus

- **neck part:** C6 – Th1, ventrally connected with larynx and trachea, laterally are lobes of thyroid gland and great neck vessels and nerves, dorsally is neck spine
- **thoracic part:** is the longest. It is in mediastinum – Th1-Th7-8. In ventral mediastinum is trachea with bifurcation, heart and great vessels, in dorsal mediastinum are nn.vagi, vv.azygos and hemiazygos, sympaticus and ductus thoracicus. Relation of esophagus to the vessels, aorta, pericardium and pleura is different according to the part of esophagus, so in case of perforation are symptoms very variable.
- **Abdominal part:** is the shortest. Hiatus esophageus is round and is in the level of Th 9-11

Inervation of esophagus

- nn.recurrentis in neck part
- branches of neck and thoracic sympaticus and nn.vagi in thoracic and abdominal part. N.X causes dilatation and sympaticus constriction of esophageal openings. Caudal part of esophagus and cardia have cholinergic and adrenergic receptors, which controls opening and closing of cardia.

Clinical physiology of esophagus

- food intake – food is moved by swallowing reflex into esophagus. Esophageal wall is very elastic and food movement is maintained by active contractions of muscles.

Examination methods of esophagus

Imaging methods

X-ray:

- normal picture is used for diagnostics of foreign bodies
- contrast examination: swallowing act with baryum paste, iodine contrast fluids if we suspect perforation
- CT and MRI in case of expansive lesions

Esophagoscopy: endoscopic examination with rigid or flexible optics

- rigid: especially for extraction of foreign bodies
- flexible especially for diagnostics

pH-metry:

by sound for diagnostics of GERD

Congenital defects of esophagus

Content

1. Aplasia, strictions
2. Tracheoesophageal fistula (TEF)
3. Congenital anomalies of great vessels
4. Hiatal hernia
5. Achalasia
6. Double esophagus
7. Brachyesophagus

Aplasia, strictions

They are caused by esophageal recanalization failure during evolvement. In this case newborn cannot swallow neither saliva nor food. Vomiting is present immediately after birth. Prognosis depends on other associated anomalies. Esophageal strictures cause dysfagia after change to solid food. Dysphagia and regurgitation need careful examination – suspicion of combined anomalies.

Diagnostics: X-ray possibly with contrast medium (iodine), CT, MR, esophagoscopy, bronchoscopy.

Tracheoesophageal fistula (TEF)

Etiology: according to Denker it is caused by failure in necking process and in forming the transesophageal septum.

Classification according to Vogt and Haight:

- Atresia without TEF
- Atresia with TEF in proximal segment
- Atresia with TEF in distal segment
- Atresia with TEF in both segments
- TEF without atresia

Symptoms: The most frequent type (88%) is esophageal atresia with fistula to trachea in distal segment. For atresia with fistula are typical: presence of spumous phlegm in mouth and nose which persist even after sucking off, vomit is without hydrochloric acid, seizures of dyspnoe and cyanosis,

asphyxia during feeding. Sometimes is present notable meteorism. In obstetric history can be hydramnion. In the most frequent type is present aeriform filling of stomach and guts.

Diagnostics: X-ray possibly with contrast medium (iodine), CT, MR, esophagoscopy, bronchoscopy.

Therapy: surgery

Congenital anomalies of great vessels

can cause esophageal patency defects as well. Most frequent are:

- Dysphagia lusoria – caused by abnormally situated a.sublclavia dextra
- Double aortal arch
- Right sided aortal arch

Diagnostics: cardiological examination

Hiatal hernia

Probably it is esophageal evolvement defect, developed around 4th embryonic week. Esophagus has insufficient length and fixation on hiatus oesophagicus, which leads to hiatal hernia.

Achalasia

Definition: it is a syndrome of non-organic cardia obstruction combined with great esophageal hypertrophy and dilatation.

Etiology: it is either improper evolvement or atrophy of plexus Auerbachi in muscles.

Symptoms: Parasympathic filament dysfunction leads to prevalence of sympathicus and sphincter contracture of cardia follows. It is presented either from birth or later, after change to solid food – vomiting after several bites, or later vomiting even predigested food. Stridor is caused by pressure on bronchus.

Diagnostics: X-ray examination of esophageal passage (saccifrom esofageal dilatation), esophagoscopy – if organic changes aren´t found, neurogenic cause of dysphagia is suspected.

Therapy: dilatation, cardiomyotomy according to Heller

Double esophagus

is rare congenital defect, when esophagus is divided by longitudinal septum. Doubling have not to go all the way throught esophagus, causing troubles and generally is found by chance.

Therapy: surgery

Brachyesophagus

Short swallow. Part of stomach lies above diaphragm, so the stomach is hold in diaphragm. Sometimes it haven´t to cause troubles, but usually it causes vomiting after every meal. Vomiting stops in upright body position.

Diagnostics: X-ray examination with contrast medium

Therapy: surgery

Inflammations of esophagus

Content:

1. Acute non-specific esophagitis

2. Mycotic esophagitis

3. Specific esophagitis

4. Stagnation esophagitis

5. GERD (gastroesophageal reflux disease)

6. Peptic esophageal ulcer

7. Sideropenic dysphagia

Acute non-specific esophagitis

Pathogenesis:

- Catarrhal inflammation of esophagus can develop during quinsy, esophageal, stomach or lung inflammation. Inflammation can spread on esophagus per continuitatem or via blood. Scarlet fever can be accopanied with ulcerous esophageal inflammation. Upper third of esophagus can be afflicetd by pseudomembranes during diphteria. Acute esophagitis can be caused by GERD, iritating food or after surgery in anaesthesia as a result of postoperative vomiting.
- Acute suppurating inflammation is localizated around decayed tumors and specific inflammations. Injury with foreign body or esophagoscopic tubus can cause this inflammation as well. Can be unconfined (flegmóna), or limited (absces of esophageal wall or periesophageal space)

Symptoms: dysphagia, odynophagia, sense of burning and chest pain

Complications: mediastinitis

Mycotic esophagitis

Pathogenesis: primary mycotic esophagitis is very rare. Yeast growth is usually caused by another condition: malignant tumor, food stagnation in esophagus, ATB or corticosteroids administration.

Etiology: most frequent is Candida albicans, less frequent Candida tropicalis, pseudotropicalis, Krusei.

Symptoms: chest pain and burning, worsening during swallowing, drought in neck and dysphagia. Suddenly arised problems are typical by patients with grave disease.

Esophagoscopy:

- catarrhal form: mucosa is red, dry and fragile
- coating form – mucosa is red, dry, hypertrophic, here and there with white coats
- pseudotumorous form
- „cotton-wool" esophagus – interior is stuffed with white mycotic bunches

Therapy: 5-10% kalium iodatum, gentian violet, antimycotics. We must be in search of basic disease, correct esophageal passage, if possible discontinue ATB and corticosteroids.

Specific espohagitis

They are very rare.

- Tuberculosis spreads in esophagus per continuitatem from mediastinal lymphatic nodes. Possible is infection via swallowed sputum or via blood.
  Symptoms: dysphagia, odynophagia, fever, malnutrition. Patients have spontanneous pain worsening during idly swallowing.

  Diagnostics: esophagoscopy – ulcer, granulations, infiltration, fistula or tuberculoma, after heal are present scars.

  Therapy: antituberculotic drugs

- Syphillis of esophagus – during 3rd stadium we can see gumma. Dysphagia is present, but not very serious

Stagnation esophagitis

Pathogenesis: food stagnate above cicatricial and functional strictures, interior of esophagus gets wider. Sometimes develop cystic esophagitis, which is caused by inflammatory infitration of mucosal and submucosal lymphatic nodes and by retention in slime glands with cyst forming.

Symptoms: chest pain and burning, dysphagia related to primary disease

Esophagoscopy: mucosal hypertrofy, bleeding, ulcers or grey-whitisch leucoplakias, irregulary granular surface of red hypertrofic mucosa

Therapy: should be focused on primary disease, temporary relief can be achieved by succking off the stagnating food, in acute exacerbation is needed local antibiotics.

Gastroesophageal reflux disease - GERD

Pathogenesis: If closure mechanism coordination of lower third of esophagus is damaged (after surgery, by hiatal hernia, etc.), gastric juices reflux to the esophagus is present.

Classification (according to Škeřík):

- catarrhal esofagitis – mucosa in lower third of esophagus is congested and swollen
- hypertrophic esophagitis – changes are primary in lower third of espohagus, but they can afflict whole esophagus
- ulcerous esophagitis – in lowr third of esophagus are areal ulcers with low granulations and fibrine coating
- fibrostenosing esophagitis – concentric strictures develop because of scars formation as a result of inflammation

Symptoms: burning, eructation, sometimes even gasric juices regurgitation, worsening in forward bend or in lying position.During swallowing can be present painful chest opression, sometimes even spontaneous chest and epigastric pain.

Diagnostics: esophagoscopy, pH-metry

Therapy: causal, diet, antacids. In fibrostenosing form is indicated sonde dilatation

Peptic esophageal ulcer

Some consider it a single nosologic unit, others consider it a quantitative change of ulcerous reflux esophagitis. It is located in lower third of esophagus, more often in men. Symptoms are more expressive then in reflux esophagitis, there can be serious hemoragies and even perforation of ulcer.

Therapy: same as in case of gastric ulcer

Sideropenic dysphagia

This disease is in literature known under those names: syndrome Gougerot-Sjógren, sy Plummer-Vinson, sy Kelly-Patterson, glossopharyngopathia sideropenica, epitheloxerosis. Almost exclusively are afflicted women.

Symptoms: dysphagia, glosodynia, mucosal atrophia of tongue and pharynx, stomatitis angularis, small mouth, dry skin, hair loss, defect nails, teeth loss, fluor albus, achylia resistant on histamin, hypochromnic anemia.

Esophagoscopy: atrophic, smoothed, glossy mucosa, white leucoplakias, semilunar folds usually from ventral side of esophagus (on X-ray looks like plain notches in contrast filling).

Therapy: vitamins B, iron by anemia, hydrochloric acid and pepsin by achlorhydria. In case of semilunar folds use spasmolytics and dilatation

Tumors of esophagus

Benign tumors of esophagus: tumors grow from mucosa or esophageal wall, they can be located even outside wall and cause oppression from outside.

Polypi: are located most frequently in upper part of esophagus or in hypopharynx, usually in men. They have gray or red colour, consistency is semi-solid, shape is long and thin.

Cysts: are multiple, of inflammatory origin or congenital cyst localizated usually in lower part of esophagus between mucosa and muscles.

Lipoma, fibrolipoma, fibroma: are usually pedunculated, so that during vomiting can be tumor vomited out of esophagus to the mouth.

Angioma: is often source of bleeding.

Papiloma: is usually around esophageal sphincters, have typical looks – pale surface, typical shape.

Adenoma: is very rare, usually is in distal part of esophagus and around cardia

Myoma: rhabdomyomas are very rare, but leiomyoma are very often, they are found in more then half cases of benign tumors in esophagus. They bulge to the interior of esophagus and are covered with normal mucosa. However, circural strictures of esophagus are also known, as well as multiple myomas and diffuse myomatosis of esophagus.

Symptoms: depends on size, shape and localization of tumor

- small tumors haven´t to cause any problems, they can be cause of spasms, transient dysphagia, feeling of opression or presence of foreign body
- larger tumors are accompanied with bigger problems with swallowing, regurgitation or bleeding if surface necrosis is present.
- Pedunculated tumors and polypi cause swallowing problems, and in case of displacement to larynx cause dyspnoe and even suffocation.

Therapy: smaller and pedunculated tumors can be removed by endoscopic nippers, loop, cauterization, laser or cryosurgically. Larger and widely nasedající tumors must be removed from external acces.

Malignant tumors of esophagus

Carcinoma is the mos frequent malignant tumor in esophagus. Sarcoma and other malignant tumors are rare. Esophageal cancer is relativelly common, in statistics of malignant tumor is on 4th or 5th place. Incidence vary depending on the area. More often are present in men (4-11:1). Carcinoma is more frequent in upper two thirds of esophagus, adenocarcinoma in lower third. Carcinoma is often result of burning of esophagus.

Classification (acording to esophagoscopic findings):

- exophytic form (nodulated shape, here and there with necrosis)

- infiltrative form (wall of esophagus is infiltrated, has limited movement, mucosa is usually not afflicted)
- ulcerous form

Symptoms: depends on localization, size and growth of tumor. Tumors with exophytic growth cause symptoms earlier. Problems in esophageal tumors are worsened by transitory spasms of esophagus around tumor, but even by reflex spasms on further places. Paralysis of n. laryngeus reccurens occur sometimes, which results in hoarseness. In later stages can develop esophagotracheal or esophagobronchial fistula.

- Tumors in area of upper esophageal sphincter cause at first paresthesia in neck, later cause dysphagia, pain, weight loss, foetor ex ore, increased salivation. In later stages can be present aspiration of food and fluids.
- Tumors in pectoral part of esophagus caouse at first painless dysphagia which is getting worse, patients have to drink during eating and finally occurs regurgitation of food during eating.
- Tumors in cardia cause dyspeptic problems, vomiting, weight loss, epigastric pain, dysphagia.

Therapy: surgery, actinotherapy. Paliative procedures are used for maintaining passage thorough esophagus – artificial endoprothesis implantation, argon or neodyn laser.

Injuries of esophagus

Burning of esophagus

Etiology:

- Alkali: receives free proton (H+) – causes majority of esophageal injuries. It causes colliquative necrosis. Typical are lyes (NaOH, KOH, Ca(OH)2), ammoniac, silicates and carbonates – contained in cleaning agents
- Acids: release proton, cause about 15% of burning injuries, especially in suicide. They are contained especially in WC and pool cleaning agents (HCl, H2SO4, H3PO4, HNO3). They cause coagulation necrosis, which limits farther penetration and damage of muscular layer. Esophagus has slightly alcalic pH and its epitel is partially protected against acids.
- Disc bateries: usually contain concentrated solution of NaOH or KOH and cause mucosal damage in 1 hour and penetration in 4 hours (damage by alkali, pressure and electricity)
- Termal damage: hot fluids (warmed up in microwave oven), tomatoes, pizza, can cause swelling of larynx and dyspnoe, esophageal injury is rare
- Drug burning: tetracyklin or NSAIDs ca cause hemorrhagies and strictures

Pathogenesis: critical pH causing esophageal ulcers is 12.5 in alkali and 2.0 in acids. Tissue damage depends on concentration and amount of given substance. Solid substances are commonly spit up, so that damage of esophagus is rare. Besides local changes organism can be influenced generally – intoxication, shock (there is no publication about death caused by systemic toxicity of alkali). Esophagus is damaged most frequently in physiological strictures, stomach is damaged in about 80% of cases.

Classification:

- 1. degree – erythema and mucosal swelling
- 2. degree – submucosal damage, ulcer
- 3. degree – muscle layer damage – perforation

Symptoms: red mucosa of moutha nad pharynx, odynophagia, dysphagia, increased salivation, nausea, vomiting. Chest and abdomen pain can signalize possible perforation. Larynx swelling is

uncommon (dyspnoe can develop in case of larynx entrance burning). Absence of damage in oral cavity doesn´t exclude heavy distal damage.

Course of disease:

- Acute phase: damage of superficial epithel with possible deeper extension and venous thrombosis. Polynuclears and bacteria infiltrate mucosa in 48 hours. Mucosa is red or cyanotic.
- Reparative phase: after 5 days in average – granulations are forming in the edge of ulcer, deposits of colagen and fibroblasts are present
- Cicatrical phase: after 2-3 weeks. If circular damage is present, patient is threaten by strictures

Diagnostics: flexible nasopharyngolaryngoscopy, blood examination, chest X-ray . Esophagosopy after 12-48 hours after injury. More serious burning in oral cavity doesn´t mean heavy esophageal damage and vice versa. Consultation with toxicologic centre.

Therapy:

- Urgent care: neutralization and diluting, in case of 3. degree (perforation) surgery is necessary – laparotomy, gastrectomy, esophagectomy. Esophagoscopy and extraction in case of battery in esophagus.
- Intermediate care: Prednison 1mg/kg/day (must not be given in case of perforation), intravenous ATB, antireflux therapy. Nasogastric sonde for 6 weeks in case of circular damage of 2.degree or perforation.
  - 1. degree: no farther therapy is needed (small risk of esophageal stricture), after 3 weeks passge with baryum
  - 2. degree: Prednison for 4 weeks, ATB for 2 weeks, H2 blockers 2-4 weeks, after 3 weeks passage with baryum
- Later care: strictures dilatation if found in anaesthesis. This is made several times a week by anterograde or retrograde way from gastrostomy (second is safer)

Complications:

- Early: perforation and mediastinitis
- Later: cicatrical strictures of esophagus, malignant tumors as a result of burning (spinocellular carcinoma)

Foreign body in esophagus

Definition: swallowing of larger foreign body (food is less often than non-food) and embeding in physiological esophageal stricture. According to duration of foreign body in the esophagus we divide them to acute and chronic.

Classification:

- organic: coins, pins, toys, batteries
- inorganic: food (bones, gristles, meat …)

Etiopathogenesis: small children put inside their mouth many different things, parts of toys. During game, running, laugh or fright the foreign body can fall inside esophagus from mouth. Foreign bodies can be from metal, plastic or is part of food – bones, stone. Bodies can have sharp or round edges. Majority of foreign bodies stay stuck in Kilian´s sphincter – first physiological stricture of esophagus.

Esophageal varices

Varices are in 90% localized in lower third of esophagus, rarely are in upper third, exceptionally in middle third.

Pathogenesis: venous plexuses are subepithelially and submucously in esophageal wall. Periesophageal varices are found near fibrous capsula od the esophagus. If blood flow through vena portae is impaired (hepatal cirrhosis or stenosis outside liver), blood have to flow throught collateral veins in esophageal wall, which are wider and varicously changed. Very rare are idiopathic varices without portal hypertension or varices in upper part of esophagus in obstruction of v. cava by tumors in mediastinum.

Symptoms: bleeding in almost 70%. If it is small and repeating it causes melena and anemia, if it is larger it causes vomiting of blood. In vomits are no gastric juices. Patient can vomit swallowed blood as well. Esophageal varices can cause little dysphagic problems.

Diagnostics: On X-ray with contrast we can find at first irregularity of lower third of esophagus, later on are present longitudinal or circular bright areas in contrast fluid. If esophagoscopy is made we can find subepithelial varices (thin, blue strips or nodes) or submucous varices (thick, strips or nodes in same color as esophagus)

Therapy: primary disease has to be cured. We must premeatus or stop the bleeding from damaged varix. To stop the bleedind hemosytptics are used as well as Sengstaken-Blakemoore balloon sound. We can sclerotizate varices by injection of sclerotizing fluid or by laser coagulation (argon or neodyn laser)

Differential diagnostics of swallowing problems

Definition:

- dysphagia: impaired swallowing
- odynophagia: pain during swallowing

Classification:

- diseases of esophagus and pharynx
- diseases outside the swallowing pathways, which cause pressure from outside
- neurologic disorder

Swallowing problems are commonly first sign of organic damage of esophagus. At first the patient is not able to swallow solid food. In later stages is impossible to swallow spoon food or even liquids. These symptoms are typical for far gone tumors or achalazia. Next group of disease with odynophagia or dysphagia are patients with foreign body in swallowing pathways. Swallowing problems accompanied with vomiting of indigested food are signs of esophageal diverticulum.

Long lasting swallowing problems lead to significant weight loss, fatigue and nutrition problems. Swallowing problem with obstruction can be accompanied with laryngeal dysphagia, when liquid food run into airways and cause aspiration bronchopneumonia.

Comments to embryology of external neck

On both sides of pharynx are branchial evaginations and branchial arches.

Branchial arches consist of mesenchym, from outside are covered with ectoderm, from inside with entoderm.

- I. branchial arch (mandibular): forms malleolus, incus, ligamnetum mallei anterior, ligamentum sphenomandibulare, mandible, m.temporalis, m.masseter, m.Pterygoideus medialis and lateralis, m.mylohyoideus, m.tensor tympani, m.tensor veli palatini, m.digastricus (ventral part) – those muscles are innervated by n.mandibularis (3.branch of n.V), as well as skin of mandible and frontal 2/3 of tongue.

- II.branchial arch (hyoid): forms stapes, processus styloideus ossis temporalis, lig. stylohyoideum, upper part and cornu minus ossis hyoidei, m. stapedius, m. stylohyoideus, m. digastricum (dorsal part), mimic muscles (m.buccinator, m. auricularis, m. orbicularis oris, m. orbicularis oculi, m. occipitofrontalis) – innervation from n. VII.

- III. branchial arch: lower part of body and cornu minus os hyoideum, m. stylopharyngeus, m. constrictor pharyngis superior – innervation of muscles and tongue´s root – n.IX.

- IV. + VI. (or + V.rudimental) branchial arch: cartilages of larynx (cartilago thyroidea, c. cricoidea, c. arytenoidea, c. corniculata, c. cuneiformis), pharyngeal and laryngeal muscles are innervated from n.X (n.laryngeus superior and inferior)

Branchial evaginations:

- I.branchial evagination (pharyngeal): lies between I.and II.branchial arch. Ventral part obliterates, dorsal part dilatates into recessus tubotympanicus. It is located between Meckel´s and Reichert´s cartilage. Eardrum, middle ear cavity and hearing tube is formed from recessus tubotympanicus

- II.branchial evagination: form fossae tonsillares and tonsillae palatinae

- III.branchial evagination: medial part forms ductus thymopharyngicus, which later perishes. Lateral part forms thymus and glandula parathyreoidea inferior

- IV.branchial evagination: medial part forms ductus pharyngobranchialis, which later perishes. Lateral part forms ultimobranchial body and glandula parathyreoidea. Base of upper parathyreoid gland descends more slowly than base of inferior parathyreoid gland from 3.branchial evagination.

- V.branchial evagination: is rudimental, forms ultimobranchial body.

Thyroid gland is unpaired organ communicating with pharnygeal cavity by ductus thyreoglossus. There is tuberculum impair in the opening. Baseof thyroid gland relatively descends into frontal aprt of neck because of embryo growth. Connection with tongue is getting thin to ductus thyreoglossus and enters lingual surface in form of foramen caecum. Ductus thyreoglossus usually totally perishes; if some remnants remain, they change into small accesory thyroid glands. It´s lower part forms lobus pyramidalis of thyroid gland

Clinical anatomy of external neck

Content

Clinical anatomy of external neck

Neck is crannialy delimited by mandible, apex of processus mastoideus and skull base. Lower border is in the level of jugulum, clavicles and C7. Important orientation points are mm.sternocleidomastoidei, hyoid bone and thyroid and annular cartilage. Under the skin is m.platysma and neck fascia. Fascia forms wrapping of muscles and nerve-vessel bunches. It has 3 parts:

- **superficialis**
- **media** – covers thyroid gland, trachea and esophagus
- **profunda** – has two leafs – alar and prevertebral. In prevertebral space isn´t any anatomic obstruction, so that pathology can spread into mediastinum

Vessels of neck

- **arteries**: a.carrotis communis divides into a.carotis interna and externa. ACI has no branches on the neck. ACE divides into a.thyreoidea sup., a.lingualis, a. facialis, a. pharyngica ascendens, a. occipitalis, a. auricularis post., a. maxillaris a a. temporalis superficialis
- **veins:** blood is drained through v.jugularis from v.jugularis interna, externa and anterior
- **lymphatic system** of neck is formed by approx 200 lymphatic nodes connected with lymphatic veins. Lymph is drained on the left through ductus thoracicus.

Innervation of neck

- motoric - n. accesorius, n.hypoglossus, n.ansa cervicalis, n. phrenicus

- autonomous - N. vagus, n.glossopharyngeus a část n. accesorius
- sensitive - n. auricularis magnus, n.occipitalis major a minor, n. transversus colli

Physiology of external neck

- there is larynx and trachea in the neck area. In the larynx is voice system.
- there is hypopharynx and upper part of esophagus as well
- in the ventral part of neck is thyroid gland with its endocrine function. It´s cells produce thyreoidal hormons – thyroxin, trijodthyronin and calcitonin. In lower part of thyroid gland are parathyroid glands, which produce parathormon. All these hormons play important role in growth and metabolism of whole organism.
- on lateral sides are important nerve-vessel bunch. In sinus caroticum are chemoreceptors and baroreceptors.
- Rich system of lymphatic vessels and nodes is connected with lymphoepithelial organs in oro- and mesopharynx. It plays important role in defense of organism.

Examination methods of external neck

- **history** – in children is acquired from parents, in older children directly from the patient. We are searching for local and global symptoms. We ask for temperature, pain, breathing, swallowing, increased salivation, changes in food intake. We ask for epidemiologic history as well – if we suspect infectious origin of disease
- **aspection and palpation** – patient´s head should be slightly bent forward. We assess symmetry of neck, color and status of skin, vescularization, edema, infiltrates, fistulae and ulcers. Palpation is made bimanually down from top – from submandibular area to supraclavicular fossa, we assess area in front of and behind mm.sternocleidomastoidei – lymphatic nodes or other formations on the neck. We assess size, shape, mobility in relation to surrounding tissues, painfullness and temperature of skin. We also examine thyroid gland.
- Laboratory examination – especially inflammatory parameters – CRP, FW, leucocytes, antibodies against EB virus, etc.
- Imaging methods: USG of neck, X-ray with or without contrast. CT and MRI require patient´s cooperation – this examination in young children has to be made in general anaesthesia. Angiography can be made for diagnostics of vascular tumors or vascular anomalies.

Examination of thyroid gland

- aspection, palpation
- imaging methods

    - USG
    - scintigraphy
    - CT, MRI
- laboratory tests:
    - T3, T4, TSH, TRH, thyreoglobulin
- examination of Achilleus ligament reflex

Congenital malformation of external neck

Content

1. Torticollis spastica
2. Fistula colli lateralis
3. Cystis colli lateralis
4. Cystis colli medialis

Congenital malformation of neck can be related to bone development – neck spondyls, which are rare. These problems belong to special deforming syndrome, which contains many congenital defects including skeleton.

Torticollis spastica

Definition: it is shortening of musculus sternocleidomastoideus on one side.

Pathogenesis: can be caused by wrong intrauterine presentation, or there can be bleeding into the muscle during birth, which cause partial fibrous transformation.

Symptoms: Child has typical head position – occiput is turned to afflicted side, face to the healthy side. The muscle is shortened and rough.

Diagnostics: history, clinical examination, ultrasonography

Therapy: Rehabilitation is successful in 90% of cases. If it is not, surgery is necessary.

Fistula colli lateralis

Definition: crevicular opening of fistula the lateral side of neck skin in front of m. sternocleidomastoideus. Fistula can be with succus or empty. Fistula can be total (has inner and

outer orifice) or subtotal. They can be on one side (more often) or bilateral. They are seen immediately after birth.

Etiopthogenesis: branchial arches are not obliterated during embryonal growth

- 1. branchial arch: coloaural fistula, its inner orifice is in the lower frontal wall of ear canal
- 2. branchial arch: inner orifice is in upper part of dorsal palatine arch – arcus palatopharyngeus. This type of lateral neck fistula is the most frequent
- 3. branchial arch inner orifice is in sinus piriformis
- 4. branchial arch: inner orifice is in upper part of esophagus

Symptoms: orifice on skin can be seen as a small dot. If the fistula has anything inside, surrounding tissue can be inflammatory changed. Fistula can be without any clinical symptoms.

Diagnostics: history, clinical examination. Fistula can be sonded, or fistulography with contrast fluid can be made.

If the fistula is inflamed and with secretion, is suitable to remove it surgically. If the fistula is whole, necessary is to remove whole fistula. In case of fistula from 2.branchial arch, necessary is unilateral tonsillectomy.

Cystis colli lateralis

Definition: painless, spherical or ovoid, circumscribed, soft tumor on lateral side of the neck, most frequently in its upper part. We can find it in any period of life, most frequently during adolescence and young adults. Usually is unilateral. The cyst contains usually mucus.

Etiopathogenesis: origin is unknown. There are 2 theories:

- Branchiogennous
- Lymphatic node origin

Symptoms: cyst can be found in any age. If it is not inflamed, it doesn´t cause any problem. If it is inflamed, it grows. We can feel fluctuation during palpation. Skin above cyst isn´t usually inflamed.

Diagnostics: ultrasonography of neck, CT or MRI. Diagnostics is confirmed by histological examination of the tissue after surgical removing. Punction is not usually made.

Therapy: surgical – total exstirpation of the cyst.

Cystis colli media

Definition: soft, spherical tumor in the medial line of neck in the level of hyoid bone. It develops, if ductus thyreoglossus is not closed. It can be seen in any age, most frequently after inflammation.

Etiopathogenesis: it develops if during thyroid gland development is not closed ductus thyreoglossus. During embryogenesis thyroid gland is formed in the root of the tongue and descends to its place. Anywhere in the course can develop medial neck cyst. According to location we can

divide cysts into: suprahyoid, infrahyoid and cysts in the level of hyoid bone. Opened ductus leads to foramen caecum. If it becomes inflamed, cyst develops.

Clinical signs: soft, not always circumscribed tumor in the level of thyoid bone. In case of inflammation skin above cyst can be inflamed as well. During palpation we can feel fluctuation. If the cyst is perforated during inflammation, medial neck fistula is formed. Pain, pressure, fever can be present during inflammation. The cyst can contain the only functional thyroid gland!

Diagnostics: clinical examination – corresponding movement of the cyst during swallowing or sticking out the tongue, ultrasonography of the thyroid gland.

Differential diagnostics: epidermoid cyst (atherom), dermoid cyst, thyroid gland cyst, tumor. Diagnosis is confirmed by histological examination.

Therapy: surgical – cyst exstirpation including hyoid bone body (operation sec. Sistrunk). If the hyoid bone body is not removed, cysts often appear again.

Inflammations of external neck

Content:

Lymphonoditis colli non specifica acuta

Definition: enlarged, painful solitary lymphatic node or lymphatic packet on the neck often with skin reaction, it tends to lymphatic node colliquation. Global state of the patient can be altered.

Etiology: most frequently bacterial origin – Staphyllococcus aureus. It not colliquate if it follows viral disease (rubella, measles).

Pathogenesis: lymphatic nodes are inflamed during or after acute inflammatory disease in the nasopharynx or oropharynx. Nodes on neck are locations where infection descends from given area.

Symptoms: painful enlarged nodes, often surrounding tissues infiltration, change of color and temperature of the skin. During palpation we can feel fluctuation – sign of suppuration. Global signs are present – fever, weakness.

Diagnostics: clinical examination, blood tests – increased inflammatory signs (increased FW, CRP, leucocytes). On ultrasonography we can see change of size, blood perfusion, signs of lymphatic nodes suppuration

Therapy: antibiotics (parenteral application is the best), local lining. Incision and drainage of lymphatic node in case of colliquation.

Lymphonoditis colli non specifica chronic

Definition: enlarged solitary node or packet of nodes as a reaction on chronic inflammation in nasopharynx or mesopharynx. Node is without signs of acute inflammation

Etiopathogenesis: nodes enlarge gradually, they are spádové nodes from chronic infection place during chronic tonsillitis, epipharyngitis or rhinosinusitis.

Symptoms: node is solid, painless, circumscribed, not fixated to the surrounding, global state isn´t changed. Chronic infection in mesopharynx, nasopharynx or in paranasal sinuses.

Diagnostics: clinical examination, blood test – increased FW, during chronic tonsillitis is increased ASLO, positive swabs from tonsils and nose, X-ray finding in paranasal sinuses (PND). Ultrasonography of neck: nodes are without shape changes, they are circumscribed, without colliquation. Differentiation from malignant affliction of lymphatic nodes is necessary.

Therapy: we must exterminate chronic infection in tonsils or nasopharynx – tonsillectomy or adenotomy.

Specific inflammation of lymphatic nodes

Definition: lymphatic nodes are solid, they are forming packets or even infiltrates, tend to colliquation and fistula formation. Treatment is difficult, they often need surgical intervention. According to origin we divide them into TBC lymphoma, actinomycosis and zoonoses.

Tuberculosis colli

Etiology: Mycobacterium avium or M. bovis

Pathogenesis: after some time lymphatic nodes packets are forming on the neck and they don´t respond to common antibiotic treatment.

Symptoms: typical place on neck is upper and middle part. Nodes form packets which merge, suppuration often appears and fistulas with secretion are formed. Nodes are palpably painful, temperature is normal or only slightly increased. Disease is now rare.

Diagnostics: epidemiologic history, ultrasonography, X-ray of chest, serologic screening, tubeculine test, microbiological examination of pus. Etiological agens is often known only after histological examination of extirpated node.

Therapy: surgical treatment and specific antituberculotic therapy

Actinomycosis colli

Etiology: etiological agent is fungus Actinomyces izraeli. It can be found in the mouth, works as a saprophyte. If patient is weakened, it can cause disease.

Symptoms: solid nodes on neck, which often colliquate and form fistulas, they are painless. Thick pus with yellow granules (clusters) flows from fistulas.

Diagnostics: clinical examination, microbiological examination with proof of clusters, eventually histological examination

Therapy: surgical intervention and specific antimycotic therapy

Toxoplasmosis

Etiology: etiological agent is protozoan Toxoplasma gonidii, host is commonly cat or another domestic animal.

Pathogenesis: transfer happen most frequently by alimentary way from insufficiently thermally prepared meat. Another possibility is direct contact – skin damage by infected animal. Transplacentar transfer from mother to child exists as well and causes congenital form of toxoplasmosis. It causes granulomatous infection of neck or suboccipital lymphatic nodes. Infection hasn´t to be acute, in 10-25% is chronic.

Symptoms: increased temperature, weakness, solid lymphatic nodes, possible colliquation in nodes.

Diagnostics: serological examination – antibody proof, specific tests, histology – granulomatous absceding lymphadenitis.

Therapy: on infectious departments. Combination of chemotherapy and ATB is given, possibly with surgical intervention – incision, drainage of colliquated node, or exstirpation.

Tularemia

Etiology: etiological agent is Francisella tularensis, transferring agent are rodent.

Pathogenesis: infection can enter through skin injury, by eating the infected meat or throught respiratory airways if contaminated dust is inhaled. Infection has several forms: dermal, pulmonary, nodal.

Symptoms: septic temperatures, flu signs, possibly necrotic tonsillitis, swollen submandibular or neck lymphatic nodes. Nodes tend to colliquation, they can form fistulas.

Diagnostics: epidemiologic history, serology – antibodies, skin test

Therapy: antibiotic combination, or surgical intervention

Bartonelosis

Etiology: etiological agent is Bartonella henselae

Pathogenesis: infection enters through skin injury after cat scratch or bite ("Cat scratch disease").

Symptoms: red pustule forms in the damaged skin, which disappears in two days. After days, weeks or even months after infection regional neck lymfadenopathy develops. Nodes are painful and tend to colliquation. Other symptoms are fever, weakness, dysorexia.

Diagnostics: serology – titre of antibodies, microbiological examination of pus – difficult cultivation

Therapy: antibiotic combination, possibly surgical intervention

Phlegmona colli, abscessus parapharyngealis

Definition: bacterial infection of deep tissues of neck with possible abscessus fromation behind fascia and around great vessels.

Etiology: bacterial, aerobs or anaearobs

Pathogenesis: infection is spreading from primary focus (suppurative processes in oral cavity, dental inflammations or pharyngeal inflammations – tonsilitis, etc.) via blood, lymph or per continuitatem. There is certain time between onset and primary infection.

Clinical symptoms: exhaustion, high and septic temperatures, antalgci head position, painfull swallowing, trismus. We can sse painfull bulge on the neck (fluctuation in case of abscess) or bulge of pharyngeal lateral wall.

Diagnostics: ENT, infectious

Imaging methods: USG of neck, examination of v.jugularis patency – Doppler, CT, MRI)

Laboratory tests: CRP, FW, leucocytes, anemia, coagulation

Microbiological examination

Therapy: parenteral ATB of wide spectrum, incision, drainage – according to localization by external way ro from pharynx. Antithrombotic therapy in case of thromboflebitis.

Complications: mediastinitis, thrombophlebitis, intracranial infections

Tumors of external neck

Content

Morbus Hodgkin – MH

Etiology: is not known. Genetics or infection with Epstein-Barr´s virus can play certain role. Disease appears especially in young people in age between 15 and 30. Majority of afflicted people are boys (75%).

Pathogenesis: lymphatic nodes affliction. Extranodal infiltration afflicts most frequently liver and spleen.

Classification:

- With majority of lymphocytes (15%) - the best prognosis
- Nodulary sclerotic type (50%)
- With mixed cellularity (30%)
- With lymphocyte depletion (5%) – the worst prognosis

Symptoms: lymphadenopathy – especially in neck and mediastinum – lymphatic nodes are solid, painless. They can merge into packets, skin above them is not changed, splenomegaly or hepatomegaly is present. If mediastinal nodes are afflicted, cough and dyspnoea can appear. Nonspecific signs: fever, night sweat, weight loss, skin itching, painfulness of lymphatic nodes after alcohol consumption.

Staging: patients are divided into 4 clinical stages according to localization and affliction extent. **Therapy** is determined according to that stage.

- I.: 1 area – lymphatic nodes or organ
- II.: 2 areas – lymphatic nodes or organ on the same side of diaphragm
- III.: affliction on both sides of the diaphragm
- IV.: diffuse affliction of organs and lymphatic nodes

Diagnostics:

- Clinical examination (e.g. ENT, oncology,…)
- blood tests – nonspecific are: increased FW, Mg, Zn, Fe, LD, ferritine, imunoglobulins
- imaging methods: ultrasonography of the neck and abdomen, CT of lungs and abdomen, skeletal scintigraphy, PET
- biopsy: histology (Reed-Sternberg cells), cytochemistry, cytogenetics, imunology

Therapy: chemotherapy (ABVD – adriamycine, bleomycine, vincristine, dacarbazine), radiotherapy up to 2 Gy. Surgical treatment is of limited importance. Radical intervention is made if it is possible to remove focus in the first clinical stage.

Prognosis: 5 years survival is up to 90%. There is high risk of secondary malignant tumors (leukemia, sarcoma, carcinoma).

Non-Hodgkin lymphoma (NHL)

Definition: heterogenic disease afflicting lymphatic tissue of neck, supraclavicular and axillar area, mediastinum, abdomen, nasopharynx, orbits and Waldayer´s lymphatic circle. NHL grow fastest of tumors in children.

Kiel classification:

- Burkitt´s type (50%)
- T-lymphoblastic type (30%)
- Large-cell NHL (20%)

Etiopathogenesis: higher risk is in children with congenital or gained immunodeficiency. Possible coincidence is with EB virus. Chronic stimulation of immune system with other microorganisms influences some types of NHL. Peak distribution of disease is in 10 years of age, boys are more often afflicted (75%)

Symptoms: mediastinal localization is most frequent. It causes brathing problems (cough dyspnoea, hemoptysis) and dysphagia. Lymphatic nodes are painless, solid and can merge into packets. Disease can express as acute abdominal disease (abdominal lymphatic nodes affliction). Nonspecific signs: weakness, nausea, loss of appetite, vomiting, weight loss, fever.

Diagnostics:

- Clinical examination: ENT, oncology, cerebrospinal fluid examination, bone marrow biopsy
- Imaging methods: X-ray of chest, ultrasonography of neck and abdomen, CT, MR
- Biopsy: histology, cytochemistry, cytogenetics, immunology

Therapy: according to stage. Chemotherapy (including cytostatics administration to the vertebral canal) – cyclophosphamide, vincristine, prednisone. Bone marrow transplantation in case of second remission. Local radiotherapy is made for local treatment of residual tumor.

Metastases into neck lymphatic nodes

Very hard, painless, little mobile or immobile infiltrates of lymphatic nodes on neck, can be unilateral or bilateral.

Etiology: they are most frequently present in malignant affliction of head and neck by carcinoma. Often is it in adult patients. These metastases are exceptional in young and adolescents. Majority of cases are metastases of thyroid gland carcinoma. In adolescents we can find metastases of nasopharyngeal carcinoma.

Pathogenesis: from primary tumor spreads infiltration by lymphatic ways to the regional lymphatic nodes in neck.

Classification of neck lymphatic nodes:

- I. submental and submandibular
- II. upper jugular
- III. Middle jugular
- IV. Lower jugular
- V. dorsal neck triangle
- VI. Ventral neck area

Symptoms: painless and solid lymphatic nodes

Diagnostics: ENT examination, endoscopy, ultrasonography of neck, CT, MR, histology. If we can´t find primary tumor on neck, we search for tumor in distant organs (mamma, lungs, stomach)

Therapy: surgical removal of neck metastases (block dissection) is made if primary tumor is operable and no other distant metastases are present. According to extent we divide block dissections:

- Total – removing all nodes on neck (area I-IV)
- Selective – removing only those nodes, which has highest probability of affliction
- Extended – include removing of other lymphatic or non-lymphatic structures (radical block dissection includes removing of n.accesorius, v.jugularis interna and m.sternocleidomastoideus).
- Those dissections are modificated according to extent of sparing other important structures (n.accesorius, v.jugularis interna and m.sternocleidomastoideus).

Paraganglioma

Symptoms: painless, slowly growing swelling near bifurcation of a.carotis with horizontal mobility (not vertical).

Neuroblastoma

One of the most frequent malignant tumors in neonates, about 5% tumors on the neck. It is tumor from sympatic ganglia. Tumor grow rapidly and metastases soon.
Symptoms: infiltration on neck without relation to lymphatic nodes, in suckligs it metastases into liver and skin
Therapy: surgical, chemotherapy, radiotherapy

Rhabdomyosarcoma

See: Rhabdomyosarcoma in Tumors and expansive processes in children

Tumors of thyroid gland

They represent about 3% of children malignancies, they have quite good prognosis. 10% of thyroid gland tumors appear up to 21 years of age, with maximum between 15 and 19 years. Risk of development is higher after irradiation (radiotherapy, nuclear catastrophe).

Classification:

- Papillary carcinoma: over 70% of thyroid gland tumors in children (causes lymphatic metastases)
- Follicular carcinoma: up to 20% of thyroid gland tumors (causes hematogenous metastases)
- Medullar carcinoma: : up to 10% of thyroid gland tumors (causes lymphatic metastases), grows from parafollicular cells. It is part of multiple endocrine neoplazia (MEN) syndrome

MEN IIA (autosomally dominant) – pheochromocytoma, medullar carcinoma, hyperparathyreoidism, develops in 12-30 years of age

MEN IIB – Marphan syndrome, ganglioneuromatosis, pheochromocytoma, medullar carcinoma

- Anaplastic carcinoma: up to 3% of thyroid gland tumors
- Tumor from Hurtle´s cells: from inflammatory changed thyroid gland cells

Symptoms: in children – multiple painless swelling in ventral and lower part of neck, lymphadenopathy in up to 90%, which is bilateral in 30%, lung metastases are present in 20%.

Diagnostics: scintigraphy (“cold nodes” are malignant in 20%), ultrasonography, CT, MR, biopsy

Therapy: surgery, Iodine 131

Malignant tumors

Content

1. Rhabdomyosarcoma

2. Synovial sarcoma

3. Neuroblastoma

4. Fibrosarcoma

5. Primitive neuroectodermal tumor and Ewing´s sarcoma

Rhabdomyosarcoma

More frequent in children of age 4-8 years (embryonal with better prognosis) or 12-15 years (alveolar type with worse prognosis). 40% is localized on head and neck.

Classification:

- Parameningeal: extents to skull base, has greater number of local recidives, worse prognosis (nose, paranasal sinuses, nasopharynx, middle ear, mastoids, fossa pterygopalatina and infratemporalis)
- Non-parameningeal: doesn´t extent to skull base

Histopathology:

- Embryonal: most frequent on head and neck (small spherical or spinocellular cells with hyperchromafinne core). We find typical striping.
- Alveolar: has higher number of metastases (not good differentiated small spherical cells evoking pulmonary alveols)
- Pleomorfic
- Botryoide

Imunohistochemy: alfa actine, myosine, desmine, myoglobine are present in embryonal tumors

Genetics:

- Alveolar type: specific translocation of long branches of chromosomes 2 and 13
- Embryonal type: mutation of chromosome 11 – area p15

Symptoms: painless formation, proptosis in orbit, respiration obstruction, Horner´s syndrome, otorrhea, ear pain. 7% cases have metastases (lungs, bones, bone marrow) in the time of diagnosis confirmed.

Staging: most tumors are stage III in the time of diagnosis

- Stage I: localized, totally resecable, without affliction of lymphatic nodes
- Stage II: resecable :
  - 2A: localized, microscopic residuum in primary localization
  - 2B: regional, completely resecable
  - 2C: regional, lymphatic nodes affliction, microscopic residuum
- Stage III: incompletely resecable or greater residuum
- Stage IV: distant metastases

Therapy:

- Surgical excision: without removing vital structures (eye) – eliminates need of postoperative radiotherapy.
- Radiotherapy: 50 Gy minimum. Complications: face growth defects, radiation induced tumors, CNS problems
- Chemotherapy: VAC (vincristine, actinomycine D, cyclophosphamide), or adriamycine, cisplatina, etoposid

Prognosis: embryonal or nonparameningeal have better prognosis

Synovial sarcoma

Slowly growing tumor in parapharyngeal and retropharyngeal space in adolescents and older children. It is formed by fibrous stroma and glandular component. Metastases into lungs, bones and lymphatic nodes are common and demand postoperative chemotherapy.

Genetics: translocation between chromosomes X and 18 is present in 90%

Therapy: excision and radiotherapy. Chemotherapy is without effect.

Neuroblastoma

Second most frequent solid tumor in children, usually in children younger than 5 years of age.

Symptoms: painless swelling, or painful bones in case of metastases (lymphatic or bone metastases are present in 70% in the time of diagnosis). Other symptoms include Horner´s syndrome, nose obstruction and nose bleeding (estesioneuroblastoma).

Prognosis: is worsening with higher age and clinical stage.

Fibrosarcoma

Is formed by fibroblasts and collagen. Often develops as a secondary tumor induced by radiotherapy. Congenital fibrosarcoma was described in neonates.

Etiology: trisomia 8, 11, 17, 20

Therapy: surgery. In children above 10 years has worse prognosis.

Primitive neuroectodermal tumor and Ewing´s sarcoma

Primitive neuroectodermal tumor is can´t be biologically distinguished from extraoseal Ewing´s sarcoma. It is formed usually in 2.decade of life. It has familiar incidence, is highly aggressive, cause distant metastases in bones and bone marrow.

Etiology: often translocation of chromosomes 22, 11 and 12.

Symptoms: afflicts usually pelvis and long bones. Head and neck affliction can be primary or metastatic (including lymphatic nodes).

Diagnostics: imunohistological – surface antigen P30/32 mic2 or HBA71

Therapy: excision, radiotherapy, chemotherapy in case of metastases

Benign tumors of neck and head

Content

1. Choristoma

2. Hamartoma

3. Teratoma

4. Myofibromatosis in children

5. Fibrous dysplasia

6. Neurofibromatosis

7. Rhabdomyoma

8. Vascular anomalies

    8.1 Hemangioma

    8.2 Vascular malformations

Choristoma

Synonym: heterotopy, ectopy, aberrant tissue

Definition: it is a normal tissue with normal structure in abnormal localization

Symptoms: they are clinically benign. They are commonly formed by solid spherical tissue. Most frequent is salivary choristoma of middle ear. Choristoma of nerve tissue is called glioma, of adipose tissue lipoma and of teeth tissue odotoma.

Diagnostics: CT, histology

Therapy: surgical removal

Hamartoma

Definition: unorganized, non-capsulated tissue in right localization.

Etiology: familiar incidence.

Diagnostics: CT, histology, family history.

Therapy: surgical removal.

Teratoma

Definition: tumor from ectoderm, mesoderm and entoderm. It is formed by fibrous, adipose, vascular, muscle, bone, chondromatous and glandular tissues.

Symptoms: In spite of the fact, that immature tissue can have signs of malignity, it is clinically benign with great prognosis. “Hairy” polyp is typical in nasopharynx. Auro-nasopharyngeal hairy polyp is described as well.

Diagnostics: histology

Therapy: surgery

Myofibromatosis in children

Definition: rare benign proliferative process. Desmoid tumor is a term used for similar fibroblastic proliferation in older children and adults.

Symptoms: local painless swelling

Diagnostics: biopsy and electron microscopy (needed for differentiation from rhabdomyosarcoma and fibrosarcoma)

Therapy: surgical removal is made in case of dysfunction or cosmetic problem. Possible are radiotherapy and chemotherapy if surgery is impossible. There is large % of recidives.

Fibrous dysplasia

Definition: benign, slowly progressing fibrous affliction of bones of unknown etiology. It is most frequent in first and second decade. Normal sponge bone is reabsorbed and substituted with immature compact bone. On skull it afflicts usually frontal and sphenoidal bone, less frequently temporal bone (external auditory canal stenosis, retroauricural oedema, lobe dislocation, development defect of ossicles or inner ear).

Classification:

- Monostotic form: (70%) afflicts only one bone (skull affliction in 10%)
- Polyostotic form: afflicts more bones (skull affliction in 50%)
- Syndrome McCune-Albright: with extraskeletal anomalies (pigmentation, endocrinopathy, growth and sexual disorders)

Diagnostics: CT, histology, increased phosphate level

Therapy: surgical. Radiotherapy can cause malignant change!!!

Neurofibromatosis

- Type I (von Recklinghausen): 85%, incidence 1:4000 births

Etiology: in 50% new mutation, rest is autosomally dominating mutation of chromosome 17 (NF gen producing neurofibromin).

Symptoms: 1/3 patients have ENT manifestation – skin affliction (“white coffee” spots), swelling on neck, larynx, orbits and salivary glands affliction

Therapy: surgical excision

- Type II: incidence 1:50 000 births

Etiology: defect of chromosome 22

Symptoms: bilateral neurinoma of n.VIII, meningeoms, schwannoms, cataract

Therapy: surgical excision

Rhabdomyoma

- Fetal develops in toddlers
- Juvenile develops in older children

Differential diagnostics: atypical cells are found. Leiomyoma and leiomyosarcoma afflict esophagus, they are often related with Alport´s syndrome

Therapy: excision

Vascular anomalies – Mulliken and Glowacki classification (1992)

Hemangioma

Definition: hemangioma is well organized formation of lobular configuration characterized by endothelial proliferation. It slowly involves.

Etiology: hemangioma is type of unregulated angiogenesis.

Pathogenesis: hemangioms in proliferative phase are formed by fast dividing endothelium, which forms syncitial tissue with or without the lumen. They have basal membrane with multiple layers. Angiogenous peptids initiate formation of capillary plexus. This process is regulated by growth inhibitor of endothelial cells.

Symptoms: it is not present in birth. It can be described as a red spot or teleangiectasia – more frequently in girls (3:1) and white race. It grows fast in early childhood. It has fast postnatal proliferative phase during first 8-12 months followed by small regression during 5-8 years. Incidence is 23% in case of birth weight under 1000g. 80% of hemangioms is isolated, 20% is multiple.

- It is solid, red formation, which is difficult to press
- Skin is reddish or bluish
- Periorbital localization: amblyopia and diplopia, astigmatism
- Subglottis: manifestation in first 6 weeks of life, in 50% together with skin hemangioma. Stridor and long lasting laryngotracheobronchitis are developing. We usually found circular swelling in dorsal part of subglottis, which can be simply pressed.

Diagnostics:

- Histology
- Imaging methods: fast flow can be present in proliferative phase (USG, MRI, CT, angiography)

- Laboratory tests: increased levels of angiogenous peptids (basic growth factor of fibroblasts) in urine and serum in contrast with vascular anomalies

Therapy:

- Usually not needed (spontaneous regression). In improper localization (orbit, larynx, oral cavity) is therapy necessary:
- Corticosteroids: formation should be smaller in 7 days, triamcinolon – application into tumor, 40 mg (1-5x) in 4-6 weeks
- Interferon alfa if corticosteroid therapy is unsuccessful. It inhibits angiogenesis and can be used during life threatening situations as well.
- Surgery: residuum removing. Subglottic hemangioma – if obstruction is present, tracheostomy is needed in more than 20%, corticosteroids (prednisone 3 mg/kg/day) for at least 4-6 weeks. If this therapy didn´t succeed, CO2 laser can be used.

Complications:

- Kasabach-Merrit syndrome: platelets cumulation in large or expansive hemangioms cause purpura and serious bleeding from GIT or brain (1%)
- Ulcers and bleeding: sudden spot bleeding during proliferative phase is caused by basal membrane damage. Repeated bleeding is rare. Ulcers are common in hamartoms in case of secondary infection.
- Heart failure from congestion in blood circulation – multiple skin hemangioms and liver affliction

Vascular formations of head and neck

Definition: structural vascular anomaly with still endothelium

Classification:

- **Low flow:**

Capillary: are formed by ecstatic veins with thin wall

Lymphatic: contain smooth and stripped muscles, nodal lymphocyte accumulation in fibrous stroma

Venous: have thin wall with irregular islets of smooth muscle

Combined: capillary-lymphatic, or lympho-venous

- **Fast flow:**

Arterial: have soft endothelium on single layer basal membrane

Arterio-venous: are incrassate with hyperplastic fibres of smooth muscle

Symptoms: manifestation is usually in childhood. They are present from birth and are characterized by normal cell growth. They grow proportionally with child, however, they can expand after injury, infection, hormonal changes, embolia or surgical intervention.

- Capillary: skin or mucosa has color of wine and contains capillaries in upper dermis. If maxilla or mandible is afflicted, there is risk of intracranial affliction (Sturge-Weber syndrome)

- Venous: dilated or ecstatic veins, common is thrombosis with calcification (flebolite). Skin and submucosa is afflicted, especially lips, cheeks and muscles (m.masseter). It is soft compressible formation. It expands during compression of v.jugularis and during Valsalv´s maneuver.
- Lymphatic: (cystic hygroma, lymfangioma) – multiple dilated lymphatic canals of variable wall thickness. Common is bleeding into cystic spaces. They can cause macroglossia, macrocheilia, macrootia and macrodontia. They usually afflict frontal and dorsal neck triangles as a big thick-wall cyst. Hypertrophy of muscles and soft tissues is usually present. They can spontaneously enlarge during bleeding or infection.

Type I: under m.mylohyoideus (cystic hygroma)

Type II: above m.mylohyoideus – afflicts cheeks, oral cavity and tongue – they are not well differentiated.

Diagnostics: USG, CT, MRI, angiography, histology

Therapy:

- Capillary: laser, surgery
- Venous: sclerotherapy (ethanol), exstirpation or both
- Lymphatic: excision, type 1 after 1 year of age, type 2 after 5 years of age
- Fast flow: embolization

Complications:

- Venous malformations: large can be connected with DIC (disseminated intravascular coagulation)
- Low flow malformations: defects of skeleton
- Fast flow malformations: bone destruction
- Dyspnoea: needs surgery immediately or tracheotomy
- Bleeding: intubation, antibiotics, surgery
- Presence of intracranial arterio-venous malformations (Sturge-Weber syndrome)

Histiocytosis

Histiocytosis X

(Lichtenstein describes idiopathic proliferation of histiocytes in 1953) Disease afflicts histyocytes called Langerhans´s cells (LCH)

Definiton: proliferation of abnormal or pathological cummulation of normal Langerhans´s cells. Disease is not malignant in classical pathological meaning. Cells are not monoclonal and have no cellular atypies. Different organ systems are afflicted (bones, skin, lymphatic nodes, liver, spleen, lings, bone marrow, CNS). Younger age means worse prognosis.

Classification:

- Eosinophillic granuloma: isolated affliction of usually only one bone, most frequently on skull (maxilla, mandible, os temporale), but afflicted can be even long bones of extremities, pelvis, ribs, spondyls. It can be found in any age, but in 5 years of age we can see 50% and in 20 years 75%. Course is typically benign and has excellent prognosis

Symptoms: bone pain with soft tissue accumulation

- Morbus Letter-Siwe: disseminated histiocytosis in children under 3 years with multiorgan affliction. Mortality is high.

Symptoms: fever, exanthema, lymphadenopathy, heaptosplenomegaly, dyspnea, changes in blood count

- Morbus Hand-Schuller-Christian: systemic histiocytosis – clinical course between previous two types. Usually afflict children in 1-5 years. Multifocal bone lesion and diabetes insipidus with limited skin affliction, lymphatic nodes and inner organs.

Symptoms: osteolysis (in 25%), exophthalmus in case of orbit affliction and diabetes insipidus in case of hypophysis or epiphysis affliction.

Diagnostics: CT, MRI, biopsy, blood count, X-ray, lumbal puncture, bone marrow

Therapy:

- Local affliction of bones: corticosteroids (including steroid injection into tumor), surgical resection, radiotherapy (60-1000 cGy, in fractions of 200 cGy per day)
- Multiorgan affliction: radiotherapy, chemotherapy (corticosteroids, methotrexate, mercaptopurine, etoposid, vincristine, chlorambucil, cyclophosphamide), immunotherapy (interferon-alfa)

Rossai-Drofmann histiocytosis

Definition: proliferation of histiocytes with lymphadenopathy (75% head and neck)

Symptoms: fever, weight loss, increased leucocytes and FW

Diagnostics: biopsy, histiocytes doesn´t have character of Langerhans´s cells

Therapy: excision, corticosteroids, chemo and radiotherapy

Injuries of external neck

Content

1. Open injuries

2. Closed injuries

3. Foreign bodies in airways

4. Aspiration

Open injuries

Definiton: injury damaging skin integrity, penetrating into neck and according to mechanism and severity can influence vital functions of organism.

Etiology: open injuries can be caused by another person, in case of suicidal attempt, extensive injuries can be caused by car accident or by dog.

Pathogenesis: wounds can be cutting, stab, gunshot or bruised

Clinical signs: according to depth of penetration it can affect only skin and submucosis or possibly through muscles. Bleeding is always present, severity depends on damaged structures. In case of ddep wounds we can see damaged great neck vessels, larynx, upper part of trachea or esophagus.

If airways are damaged, air can get into submucous tissue and causes submucous emphysema. It causes crepitation during palpation. Other signs are dyspnea, suffocation, dysphonia, dysphagia, hemoptysis

Diagnostics: endoscopy, X-ray (we can see foreign body or air in submucous tissue), CT

Therapy: wound cleaning, stop bleeding, suture of damaged structures and skin, possibly foreign body extraction. If airways are damaged, intubation or tracheotoy is necessary

Closed injuries

Definition: integrity of skin isn´t damaged

Etiology:

- mechanic injury: blunt force, fall on handle bars, wire, car accidents, aspiration, swallowed foreign body
- termic injuries: hot steem or hot fluids
- chemical damage: inhalation or ingestion
- iatrogenous damage: by nasogastric sound or during endoscopy

Pathogenesis: in light injuries there hasn´t to be larger damage

Cartilages of pharynx and trachea in children are elastic – soft and supple. In case of greater force laryngeal skeleton can be damaged, Termic and chemical wounds cause hyperemia, edema, possibly mucosal necrosis. Necrosis causes scars.

Clinical signs: subcutanneous hematoma or excoriation after blunt damage. In larnyx: hematoma, edema, subcutaneous emphysema, mucosal damage with bleeding. General signs – dyspnea, dysphonia, irritating cough, temporary breathlessness during laryngospasm, hemoptysis. These symptoms can be present in case of chemical vapors inhalation (excluding hemoptysis). If swallowing pathways are damaged we can see dysphagia, odynophagia, increased salivation, fresh blood in salivae.

Diagnostics: history, ENT examination, endoscopy, X-ray, CT

Therapy: acording to extent – observation, antiedematous therapy, intubation or tracheotomy if injury is extensive. Endoscopy and local treatment, foreign body extraction.

Foreign bodies in swallowing pathways

see: Foreign bodies in esophagus in Injuries of esophagus

Aspiration

see: Aspiration in Injuries of larynx and trachea

Differential diagnostics of neck lymphadenitis in children

Content

Diagnostics

ENT examination:

- inspection: asymmetry, changes in vascularizetion, color, scrs, fistulae, abnormal pulsation, localization
- palpation: mobility, consistency, fluctuation, pulsation, crepitation, size
- biopsy: FNAB need cooperation!! – it is not method of choice for children. Negative result cannot be described as a absence of malignancy. Method of choice is excision or exstirpation

Imaging methods:

USG, CT, MRI, angiography, contrast examination (esophagus), scintigraphy (thyroid gland)

Other clinical examinations:

pediatric, stomatologic, ophthalmologic, infectious, surgical, ortopedic

Laboratory tests: blood test (CRP, leucocytes), antropozoonoses, bacteriology, …

Differential diagnostics

Inflammations of lymphatic nodes

Approx 40% of healthy children (up to 1 year of age) have enlarged lymphatic nodes in front of ventral edge of m.sternocleidomastoideus

Viral infections:

- reactive lymphadenitis in case of viral infection in airways, oral cavity or pharynx
- mononucleosis (EBV, CMV)
- HIV
- Herpesvirus 6
- Varicella – zoster

Bacterial infections:

- reactive lymphadenitis in case of bacterial infection in airways, oral cavity or pharynx (S. aureus, Streptococcus A)
- Mycobacteria (TBC, non-TBC)
- Antropozoonoses (brucelosis, tularemia, bartonelosis)

Other infections

- mycotic lymphadenitis
- toxoplasmosis
- actinomycosis
- chlamydia infections
- Treponema pallidum

Non-infectious affliction of lymphatic nodes:

- rheumatoid arthritis
- lupus erytematodes
- sarcoidosis
- Kawasaki syndrome
- Kikuchi-Fujimoto syndrome
- Castelman syndrome

- Rosai-Dorfman histiocytosis
- PFAPA syndrom

Inflammations out of lymphatic nodes

- inflammations of salivary glands
- phlegmons and abscesses
- infection of airways, swallowing pathways, ears, teeth, angina Ludowici
- thyreoiditis

Congenital defects and anomalies

Approx. 50% of cutted out formations

- lateral or medial cyst or fistula – 30%
- dermoid or epidermoid cyst (atheroma)
- vascular anomaly
- thyroid gland cyst
- anomaly of thymus (cystic formations espeially on the left side)
- anomaly of skeleton (Klippel-Fei sy – cervicothoracal synostosis)
- anomaly of muscles (torticollis – unilateral contracture of m.sternocleidomastoideus)
- anomaly of skin
    - pterygium colli – Turner´s sy, Noonan sy
    - middle neck cleft – abnormal merging of branchial arches
- heterotopy (choristoma)
- laryngocele
- Zenker´s diverticulum

Tumors

usually benign, in case of malignant it is usually sarcoma.

- Lymphoma
- Sarcoma
- Tumors of thyroid gland

- Tumors of slaviary glands
- Histiocytosis X
- Neurogennous tumors
- Dermal tumors
- Teratoma
- Metastases

Postoperative states, injuries

- Emphysema
- Hematoma
- Keloid scar

Differential diagnostics according to localization

The most frequent pathology in children in specific localizations. Malignant tumors asre usually behind m.sternocleidomastoideus and above clavicle. Malignant tumors are clinically commonly asymptomatic swelling, thick and painless, innervation defects are common.

Submandibular region

Inflammations:

- Lymphadenitis
- Sialoadenitis

Congenital malformations

- Vascular anomalies
- Cysts and fistulae

Tumors

- Tumors of salivary glands
- Lymphoms
- Rhabdomyosarcoma

Ventral edge of m.sternocleidomastoideus

Inflammations

- Lymphadenitis

Congenital malformations

- Cysts and fistulae
- Laryngocele
- Vascular anomalies
- Anomaly of thymus

Tumors

- Lymphoma
- Rhabdomyosarcoma
- Paraganglioma

Dorsal edge of m.sternocleidomastoideus

Inflammations

- Lymphadenitis

Congenital malformations

- Vascular anomalies

Tumors

- Lymphoma
- Metastases (nasopharyngeal carcinoma)

Supraclavicular region

Congenital malformations

- Vascular anomalies

Tumors

- Lipoma
- Lymphoma
- Metastases (from lungs, esophagus, kidneys, testes)

Anatomy, physiology and voice development

Clincal anatomy of larynx and trachea were already desribed, so we will describe here only facts about voice formation.

Structure of larynx and vocal cords were described by Hiran in 1974. Vocal cords have multiple layers.

1. On the inferior surface is pseudostratificated spinocellular epithelium. Spinocellular epithelium is also on the medial parts of vocal cords.

2. Subepithelial tissue – lamina propria has 3 layers:

    - Reincke´s space – thin tissue with small amount of fibroblasts and collagenous and elastic fibres allowing vibration of mucosa in relation to ligamentous and muscular base

    - Middle layer has especially elastic fibres and average amount of fibroblasts. They form part of ligamentum vocale. Fibroblast in this layer are responsible for cicatrication of vocal cords after phonosurgical operations or other pathology in this layer

    - Deep layer has especially collagenous fibres and the rest is ligamentum vocale

3. Last layer of vocal cords is musculus vocalis (m.thyreoarytenoideus)

Muscles of larynx are divided accordin to function:

1. Adductors – innervated by adductor brach of n.laryngeus recurrens

    - Lateral part of m.thyreoarytenoideus

    - M.cricothyreoideus lateralis

    - M.interarytenoideus – is innervated bilaterally from both n.laryngeus recurrens

2. Abductors – m.cricothyreoideus posterior – innervated by abductor branch of n.laryngeus recurrens

3. Tensors

    - M.cricothyreoideus – innervated by n.laryngeus superior

    - M.thyreoarytenoideus – innervated from n.larryngeus recurrens – antagonist of the previous muscle

Motoric fibres of n.X goes from nucelus ambiguus in prodloužená mícha, they merge together and go together through foramen jugulare. After that they divide again to innerve specific muscular groups. N. laryngeus sup.leaves under foramen jugulare, under ganglium nodosum. N.laryngeus recurrens leaves near a.subclavia l.dx.and aortal arch. It goes under esophagus and trachea and returns to larynx. This anatomic relations are substantial for diagnostics of n.larnygeus recurrens lesions. Affliction of both branches point at damage in the foramen judulare or higher. Idiopatic affliction of recurrent nerve demands detailed examination of mediastinum.

Physiology of voice

See also Physiology of larynx

Voice organs:

1. Diaphragm and lungs – generator of air flow

2. Larynx – voice generator of fundamental frequency

3. Tube – (hypo-, meso-, nasopharynx, oral and nasal cavity, sinuses) – modulator of voice

Sound is formed primary in larynx, it means increase and decrease of air pressure in larynx, which is modified in tube. Vocal cords approximate near each other and subglottic pressure is increased.This pressure progresively inreases because of diaphragm contraction and glottis is opened thanks to its elasticity. Air flow through glottis cause Bernoulli´s effect, which close glottis with cooperation with pressure decrease subglotically and pressure increase supraglotically. This procedure is repeating and sound is formed – laryngeal tone.

Vocal cords oscillate in horizontal level and mucosa is undulating vertically. Changes in mucosa elasticity or bad position of vocal cords interfere with correct oscillation of mucosa. This results in hoarseness. However, changes in supralaryngeal space can cause change of voice.

Change of voice intensity increases with increasing air flow through glottis and with decreasing vocal cords tension. Change of voice is related to vocal cords tension and voice intensity. Vocal cords tension cannot be increased without limits, however, frequency range of human voice can be much more than 2 octaves. This restrictions of human voice result from oscillation changes in only specific part of vocal cord – register:

- Thoracic – vocal cord oscillate in its whole range from comissura anterior to cartilago arytenoidea
- Middle – vocal cord oscillate only in its membraneous part from comissura anterior to cartilago arytenoidea
- Head – vocal cord oscillate only in its anterior half of membranous part

Fistule (*not fistula*) – special register chracterized by thin vocal cords (significant tension), where only edges of ventral hlaves oscillate.

Development of voice

See Comments to embryology of larynx and trachea

Voice is present in all physiologic noenates in time of birth. Length of vocal cords is 3-5 mm, and level of scream cca 400 Hz. There isn´t any intersexual difference until pubescence. Voice is continuously getting deeper in its base frequency. It is cuased by larynx growth. Length of vocal cords in 9 years old children is 6-7 mm and voice level cca 300 Hz. Significant changes happen in pubescence, when larynx rapidly grow. Time of grow is cca 6-12 months. Start is very individual, it is sooner in girls (10-18 years) and later in boys (12-20 years) and is connected with development of secondary gender signs. Frequency of voice decreases by half in boys thanks to vocal cords lenghtening to 25 mm, in girls to 15 mm. During mutation are vocal cords overvasculrized and edematous. In adult men is level

of converstion voice 80-120 Hz and in women 170-260 Hz. Range of singing voice is of course much more wider.

Other significant changes start after 6.decade of life, when atrophy of vocal cords begins, changes in respiratory system happen and there can be other diseases influencing the voice. Result is narrow frequency and dynamic range , dyspnoic voice and insignificant intersexual differences.

Examination methods of voice

Symptoms

Perception of voice quality is very subjective and there is no norm for voice. Differences depends on cultural and socioeconomic factors.

Dysphonia (hoarseness) – pathologically changed voice quality. Causes:

- Irregular vocal cords oscillation
- Change of glottis closure

Other symptoms in case of voice pathology

- Narrow dynamic range of voice
- Narrow frequency range of voice
- Increased phonation force
- Decreased maximum phonation time
- Odynophonia
- Dyspnea during phonation

Examination of voice

Perceptive examination of voice – subjective assessement of voice by examinator. Subjectivity is a problem, but it is simple and every doctor can do this. Phonation assessement according to UEP (Union of Europian Phoniatrists): 0 – normal voice, 1 – slurred voice, 2 – mild dystonia, 3 – moderate dystonia, 4 – serious dystonia, 5 – aphonia, 6 – voice loss after total laryngectomy. GRBAS scale: G-grade, R – roughness, B – breathinness, A – aesthenicity, S – strain

Assessement of phonation start – soft vs. hard or dyspnoic

Measurement of maximum phonation time

Measurement of dynamic and frequency range and assessement of voice array - dynamci range in all frequencies

Spectral analysis of voice – thanks to Fourier´s transformation we can see presence of specific frequencies in given time

Multidimensional analysis – base parameters: jitter, shimmer, harmonic, base frequency and tens of other parameters

Examination of speech organs

Electroglottography – measures changes of electric resistance caused by vocal cords oscillation recorded by electrodes put on thyreoid cartilage. It is often used for stroboscopic light synchronization istead of microphone.

Indirect larnygoscopy

Laryngostroboscopy – gold standard in examination methods for patients with voice disorders. Method has its limit in resolution of human eye, which can record maximum 20 pictures per sec.

The most deep men´s voice has cca 50 Hz, sod irect examination of vocal cords mobility is impossible by sight. Stroboscopy use blinking light. If we light regular periodic process by light flashes with same frequency as this process, it seems, that the picture is without movement (wee see vocal cords in same position). If requency is slightly increased, we can see this process slowly moving. Today larnygostroboscopes automatically synchronize their frequencies of flashes with patient´s voice according to microphone or electroglottography. Disadvantage of this examination in impossibility of stroboscopic effect in severe hoarsenesses, where vocal cords move so irregulary, that synchronization is impossible.

Videolarnygoscopy/videolaryngostroboscopy – previous examination with endoscopic camera.

Videocymography – record of very narrow part of vocal cords thanks to special high frequency camera, working at 8000 Hz, so it can display even irregular processes, or processe that last very shortly.

High speed videolaryngoscopy – endoscopic camera operating at 2000 Hz. This method has advantages of both previous methods (records iregular processes and displays vhole vocal cords)

Electromyography (EMG) – analysis of eletric activity of muscular motoric units. IT is neurologic method, but it can be made by ENT specialist skilled in this method.

Pneumography – method informing us about movements of thoracic and abdominal walls during phonation

Spirometry – informs about lungs capacity, which is one of the most significant factors of phonation time

Imaging methods, laboratory tests and other examinations are use only for specific cases.

Functional and psychogennic voice disorders

1.Voice misusing or abusing

1.1 hyperkinetic dysphonia and hyperkinetic dysphonia in children

1.2 hypokinetic dysphonia

2.Psychogenic dysphonia and voice neurosis

- 2.1 psychogenic aphonia/dysphonia
- 2.2 spastic dysphonia
- 2.3 phonastenia
- 2.4 dysphonia hysterica
- 2.5 problems in adolescence
  - 2.5.1 voxi fistulosa persistent
  - 2.5.2 mutatio prolongata
- 2.6 vox ventricularis

Voice misusing or abusing

Hyperkinetic dysphonia and hypekinetic dysphonia in children

Etiology: it is caused by excessive voice exertion, by too loud speaking or screaming. In children it can be caused by simulating voices (e.g. cars, planes, fairy creatures, etc.), which means voice use in improper position. Incidence is significantly influenced by culturally-ethnic factors (temper and communication style), by alcohol consummation and smoking.

Symptoms: in early stages: blurring and fatigue of voice, on vocal cords aren´t any changes. Doctor is attended after voice worsening into hoarseness, often with aphonic episodes. Voice is hoarse, compressed with possible dyspnoic component, with hard voice start. On neck we can see protuberant veins and clenched neck muscles. Shortened maximum phonatory time and narrowed voice spectrum. During laryngoscopy we can see fusiform thickening of vocal cords with maximum in centers (in speaking voice, however, place of maximum depends on used voice in abusing – location of maximum oscillation of vocal cords). Result is glottis insufficiency in the shape of hourglass. We can see small inflammatory changes – small swellings, venous congestion, small succus. In stroboscopic light we can see increased tension of vocal cords, reduced movement of medial edge of vocal cords and decreased oscillation amplitude, lengthen phase of closure.

Therapy:

1. basics of vocal hygiene (not scream, adequate voice level, shorten speeches, voice augmentation – microphone, alternative communication during presentations – pictures, videos)
2. reeducation of voice (methods used by author):
   a. Correct position of body (relaxed position, limbered up neck spine, etc.)
   b. Breathing exercises (accent on abdomen breathing, active breath conduction)
   c. Relaxing phonatory and articulation muscles
   d. Practice of soft start, tender amplification and modulation of voice
   e. Aplication of exercises in words and after that in sentences

3. Symptomatic therapy of laryngeal inflammation (adstringents, expectorants, in case of positive microbiological cultivation ATB locally or generally)
4. In children (until 4-7 years) and non-cooperative patients is therapy often limited to instructions to parents or guardians

Patient is followed until voice reeducation is accepted by patient, or until voice improvement. Specificity of therapy in children is too different, that in children is this diagnose separated – hyperkinetic children dysphonia

Complications: if voice use isn´t improved, patient is endangered by organic changes of vocal cords: voice node, cyst, polyp, hematoma or atrophy of cord.

Differential diagnostics: spastic dysphonia has staccato limiting communication. In case of organic changes we must exclude tumors and chronic laryngitis.

Hypokinetic dysphonia

Etiology: atrophy of m.vocalis and fibrous tissue in lamina propria of vocal cord based on voice abuse and natural degenerative processes of aging. Other diseases can participate.

Examination: voice is blurred, hoarse, with significant dyspnoic component.In laryngoscopy we can see fusiform or semilunar rima. In case of light affliction we can see only visible sulcus vocalis (sulcus in medial edge of vocal cords visible only in stroboscopic light). In stroboscopic light is seen reduced tension of vocal cords, displacement of medial edge laterally, short or incomplete phase of closure.

Therapy: basics of voice hygiene, voice reeducation, vitamins B. If therapy is unsuccessful and voice is very dyspnoic, surgical therapy is possible – injection of matter into vocal cord – it is pushed more medially so that insufficiency is reduced.

Psychogenic dysphonia and voice neurosis

Psychogenic aphonia/dysphonia

Etiology: it is conversion disorder, which is caused by psychic or emotional trauma (conversion disorder is disease simulating another somatic disease, when patient doesn´t pretend symptoms, but he believes, that he is sick)

Symptoms: voice is usually weaker or dyspnoic (changing in time), often connected with respiratory infection.

Examination: sonant voice, gag reflex. In laryngoscopy: vocal cords at first shortly adduct in middle line and after that quickly abduct into respiratory position and cause aphonia. During cough we can see good vocal cords adduction and normal mucosal movement.

Therapy: examination by doctor often reliefs from problems. If not, voice rehabilitation follows similarly like in reeducation in vocal cords movement disorders: during cough or with help of gag reflex. We must tell patient, that if he has normal voice during cough, he can have normal voice even

during normal speech. It must be noticed, that primary problem wasn´t solved and definitive resolution belong s to psychologist. If all this is unsuccessful, we can (with consent of psychologist) irritate larynx with laryngeal brush and treat patient with power.

Differential diagnostics: spasmodic abductor dysphony – affliction of pyramid track, symptoms are more constant and don´t respond to classical therapy. Now it can be treated by botulotoxin application into vocal cords (see Spastic dysphonia). If paresis of n.larryngeus recurrens and subluxation of arytenoid cartilage are present, there is abduction position of vocal cords during whole examination.

Spastic dysphonia

Etiopathogenesis: predominance of adductor muscle groups with significant psychic component. In presence appear some new knowledges. Placing into this group is historical.

Symptoms: compressed, clenched voice, which has staccato characteristics and evoke stammer. IT disappears during singing and emotional manifestations (laugh). Sometimes it is accompanied by spastic torticollis and/or spasm of facial muscles: blefarospasm, spasms of m.orbicularis oris, uncontrollable movements of lower jaw.

Diagnostics: in laryngoscopy we can see spasm of:

- vocal cords, when they are pressed together so strongly, that medial edges are elevated or only irregular oscillations are visible
- vocal cords and ventricular plicae – glottis is not visible, in laryngoscopy we can sometimes see oscillations of ventricular plicae
- supraglotic space – epiglottis is moving towards larynx interior and aryepiglotical plicae are closing – similar to laryngospasm

Larynx is moved up during voice attempt.

Therapy: voice reeducation is usually unsuccessful. Another possibility is neurotomy of n.larryngeus recurrens – unfortunately, symptoms appear again in several years. Quite good results have botulotoxin application into afflicted muscles.

Complications: organic changes on vocal cords if it lasts too long

Differential diagnostics: abductor dysphony – affliction of pyramid track. Clinical picture is same (only higher onset of other muscles spasms), it can be distinguished only according to botulotoxin effect. If the effect is immediate and maximal, it is a placebo effect and cause is psychic. In case of neurogenic cause is effect maxed after week. Other neurologic diseases have to be excluded, they usually influence speech or other motoric functions (swallowing, eyelids closing). Laryngospasm is shorter and dyspnea is present. Ventricular voice doesn´t have staccato and has effect even during singing.

Phonastenia

Etiology: is caused by excessive fear from failure during patient´s presentation (singer, actor, business-man or lawyer during meeting)

Symptoms: voice efficiency affliction – weak and easy tiredness of voice, decrease of dynamic and frequency range, tremolo (frequency fluctuation greater than 7 Hz).

Therapy: improving voice technique, psychotherapy

Differential diagnostics: exclusion of neuromuscular diseases (myasthenia gravis)

Dysphonia hysterica

Has variable character, usually seen in women. Typical is many underwent examinations and therapy methods.

Symptoms: increased voice fatigue, spastic dysphonia, other somatic symptoms: dryness in mouth, scratching in throat, lot of mucus.

Therapy: psychotherapy

Problems in adolescence

Vox fistulosa persistent

Etiology: emotional stress from psychosocial changes in adolescence

Symptoms: voice stays in inadequate high position, similar to voice of boy

Diagnostics: Appear secondary changes (often fusiform insufficiency of vocal cords) as a result of voice overburdening by inadequate voice position.

Therapy: voice reeducation, psychotherapy if needed

Complications: if it isn´t early improved, secondary changes appear like in functional disorders from abuse or misuse.

Mutatio prolongata

Etiology: same as vox fistulosa persistent, base problem is often excessive fixation of son on mother. It is often connected with growth acceleration and patient doesn´t psychically handle his somatic change.

Symptoms: voice skipping even after 18.year of age and proper growth

Therapy: voice reeducation + pressure on pomum Adami to lower the voice. It can be supplemented by demonstration of voice level with help of musical instrument, or by usage of patient´s voice record.

Vox ventricularis

Symptoms: hard, rough and deep voice with significant aspirate

Diagnostics: irregular oscillation of meatusiculat plicae in laryngoscopy, vocal cords are hidden

Therapy: voice reeducation, local anaesthesis or larynx anemization can help, because it premeatuss contact of ventricular plicae

Differential diagnostics: we must differentiate it from vocal cords scars or tumor (microlaryngoscopy if needed). Difficulties can cause differentiation from spastic dysphonia.

Speech disorders caused by mucosa and submucous tissue affliction of vocal cords

Content

1. Inflammations
    - 1.1 Acute laryngitis
    - 1.2 Chronic laryngitis
2. Organic abnormalities caused by voice abuse or misuse
    - 2.1 Noduli plicae vocalis
    - 2.2 Polypus vocalis
    - 2.3 Cystis vocalis
    - 2.4 Ulcera contagiosa plicae vocalis
    - 2.5 Haematoma plicae vocalis
    - 2.6 Sulcus vocalis

3 Other local lesions

4 Fibrosis

Organic abnormalities caused by voice abuse or misuse

From surgical aspect those diagnoses belong to chapter of organic lesions for their organic findings. From phoniatric aspect belong to chapter of functional disorders for their etiology.

Noduli plicae vocals

Etiology: hyperkinetic dysphonia. I is not clear why someone has nodes after short time and someone after longer time or even never at all. Probable cause is histological differences in vocal cords structure.

Symptoms: same as in hyperkinetic dysphonia

Diagnostics: same as in hyperkinetic dysphonia (according to voice we cannot say if nodes are present). In laryngoscopy we see limited thickness of vocal cord of same color. Most frequent onset is half or in the border between anterior and middle thirds. Other changes are same as in hyperkinetic dysphonia. In stroboscopic light we see oscillation fade-out of medial edge near node (result of cicatricial connection of mucosa and lamina propria)

Therapy:

- Conservative: - voice hygiene, reeducation and medication (adstringents, aescin, mucolytics)
- Surgical – especially in bigger nodes (after removal is necessary one week of absolute voice rest and after that voice reeducation)
- Therapy in children: up to ¾ can be cured by conservative therapy (including laryngoscopy normalization) and 10-15% can improve their voice by conservative therapy to such level, that they doesn´t need any other therapy. Surgical intervention is made only in strictly indicated cases in patients with high motivation, compliance and need of fast treatment (singer, actors). Reeducation has to always follow. In case of bad decision is patient endangered by postoperative complications.
- Complications: often recurrence or even polyp without voice reeducation (removing of hyperkinetic dysphonia). Complication of any surgical intervention on vocal cords is inflammation followed by cicatrization with catastrophic effects on voice.

Differential diagnostics: exclusion of tumors

Polypus vocalis

Etiology: bleeding into node after voice exertion. Basic is onset of dilated vessels near node.

Symptoms: voice changes depend on polyp location. If it is located on upper surface, we hear two frequencies in voice (diplophonia), if it is located into glottis – significant dyspnoic component. Other symptoms are same as in hyperkinetic dysphonia.

Diagnostics: in laryngoscopy we see polyp on vocal cord or in glottis. In front of and dehind the polyp is large insufficience causing significant dyspnoic component. In stroboscopic light we see different amplitude of vocal cords oscillation, on side with polyp we see no movement of medial edge. Definitive diagnosis is determined after histological examination.

Therapy: surgical + voice reeducation after that

Complications: often recurrence without voice reeducation (removing of hyperkinetic dysphonia). Complication of any surgical intervention on vocal cords is inflammation followed by cicatrization.

Differential diagnostics: exclusion of tumors

Cystis vocalis

Etiology: hyperkinetic dysphonia, hematoma of vocal cord

Symptoms: hyperkinetic dysphonia

Diagnostics: in laryngoscopy we ca see circumscribed formation of different color than mucosa (white or yellow). Can be bilateral or unilateral. Histology is needed for diagnostics.

Therapy: surgical + voice reeducation after that

Complications: often recurrence without voice reeducation (removing of hyperkinetic dysphonia). Complication of any surgical intervention on vocal cords is inflammation followed by cicatrization.

Differential diagnostics: exclusion of tumors

Ulcera contagiosa plicae vocalis

Etiology: injury of larynx by excessive screaming – usually in affect

Symptoms: increased voice exertion, blurred voice, cough

Diagnostics: in laryngoscopy we see whitish coat on medial parts of arytenoid cartilages, with bloodshot rim.

Therapy: voice rest, local placation of corticosteroids, or possibly local application of ATB (After cultivation)

Differential diagnostics: inflammation, tumor

Haematoma plicae vocalis

Etiology: hematoma isolated on one vocal cord is caused by voice exertion itself or during inflammation (acute or chronic laryngitis)

Symptoms: only slight changes – blurred voice, voice fatigue, limited voice range

Diagnostics: in laryngoscopy we see vocal cords with dilated vessels and hematoma of variable location and size. In stroboscopic light we see lesser oscillation or no at all of afflicted vocal cord, medial edge isn´t moving. In case of recurrences is suitable to examine coagulation and capillary fragility.

Therapy: voice rest, hemostyptics (etamsylate, vitamin K)

Complications: in case of recurrences can develop polyp or cyst

Differential diagnostics: hematoma of larynx caused by injury, tumor, venous malformation, hemangioma

Sulcus vocalis

Early phase of vocal cords atrophy – see Hypokinetic dysphonia

Other local lesions

Reincke´s oedema

Hyperkeratosis and other precancerosis (see Tumors of larynx and trachea)

Fibrosis

Etiology: reactive scar formation after injury (see Injuries of larynx and trachea), surgery or radiotherapy. Specific inflammation – Crohn´s disease, sarcoidosis, Wegener´s granulomatosis, amyloidosis or hormonal causes (hypothyreosis, hirsutism and others)

Diagnostics: vocal cord dilatation in laryngoscopy, during stroboscopy we can see decreased oscillation of vocal cord, which is caused by decreased plasticity.

Therapy: only voice rehabilitation

Differential diagnostics: tumors

Voice disorders from voice ligament affliction

See Tumors of larynx and trachea

See Injuries of larynx and trachea

Neuromuscular causes of hoarseness

Atrophy of musculus vocalis – *see hypokinetic dysphonia*

„spasmodic“ dysfony (abductor, adductor) – *see differential diagnostics Spastic dysphonia and Psychogenic aphonia/dysphonia*

Muscular weakness – muscular dystrophia, amyotrophic lateral sclerosis, poliomyelitis, bulbar and pseudobulbar unilateral paralysis, bilateral paralysis of n.laryngeus recurrens, unilateral paralysis of n.laryngeus superior, syndroms of foramen jugulare

Anatomy of speech organs

Anatomy and physiology of articulation organs (nose, oral cavity, pharynx, larynx and trachea) – see specific chapter.

Physiology and development of speech

Content

1. development of speech
    - 1.1 sound level
    - 1.2 lexical level
    - 1.3 grammar level
    - 1.4 pragmatic level
2. language levels
3. Fonetics

Development of speech

Development (ontogenesis) of speech interests doctors, psychologists, fonetics specialists and special pedagogues. Speech development goes through stages. Borders between those stages are undistinguished and everyone have to go throught these stages. Different can be only length of those stages.

Preverbal stage – gaining habits, on their base is later formed speech. These activities play very important role in this (suction, chewing, swallowing)

- first sign is scream (in first few weeks is scream short and monotonous
- after 6 week has scream emotional character, at first it means dissatisfaction – hard voice start (damages vocal cords at most)
- later (2-3 months) even satisfaction – soft voice start
- period of instinct babbling (4-6 months) – it is „game with speech organs“. Child makes same movements as during eating, followed by sound (even in deaf children)
- period of reproducing babbling (6-8 months) - sound and sight control (deaf children stop to babble)

- stage of speech understanding (8-12 months) – child didn´t understand meaning of word, but on the base of this word can watch something or someone and react on specific challenge (usually motoric reactions)
- speech development itself (1 year) – first real sgins are words, which forms sentences (with only one word), mean wishes, emotions and demands – emotionally free stage. Words have connection with specific persons and things, emotional characteristics, accent and intonation have great importance
- egocentric stage (1,5-2 years) – child imitates adults, reveals spech as a activity, repeats words itself – rapid qualitative and quantitative speech development
- development of communication speech (2-3 years) – child learns to reach variable goals
- stage of logic meanings (3 years) – abstraction
- in 3-4 years child present its thoughts quite precisely, next development is related to intellectualization of speech

Language levels

Sound level (fonetic – fonologic)

Development starts around 6-9 months – transition form instinct to reproducing babbling. In child speech are at first fixed vocals and at last typical signs of mother language. Physiological development of sound part ends around 5.year (at latest in early school age)

Lexical level (lexical-semantic)

- in 10.month starts development of passive word-stock
- in 1.year first words – hypergeneralization (haf, haf – everything, what is hairy and has four legs)
- subsequently develops hyperdifferentiation (daddy means only child´s own father)
- first age of questions - 1,5 year – „What is it?“, or „Who is it?“.
- Second age of questions – 3,5 years – „Why?“ or „When?“

Grammar level (morphologic-syntetic)

Around 1.year of age, words act as a sentences. In 1,5-2 years are „sentences“ connected toghether. Children use at first substantives and after that verbs, between them are used onomatopoic interjections, in 2-3 years use afjectives, pronouns, prepositions, conjuctions, numbers. Singular and plural after 3.year, complex sentences formation in 3-4 years of age.

Pragmatic level

According to Lechta, it is level of social aspects of communication

Fonetics

It is science about sound part of language and functions during speaking. IT contains: consonants and vocals formation, their perception and their use in language.

It describes sound from 3 aspects:

1. How is sound formed
2. What is the acousitc essence of sound
3. How is sound of speech perceived during communication

Ortophonia – science about correct formation and sounding of vocals and consonants

Ortoepia – science about correct use of speech sounds

Phonology – studies system of language sounds and its phonems

Speech sounds are divided into:

- vocals
- consonants

Differences between them:

a. articulation aspect
    i. vocals – air passes freely through resonant cavities, mouth is open
    ii. consonants – air must pass the obstruction, mouth is closed
b. acoustic aspect
    i. vocals – sounds like tones
    ii. consonants – causes murmur

Logopedic diagnostics

Logopedic diagnostics is necessary for choosing the logopedic treatment as well as for diagnostics. The most important is continuous spontanneous speech of individual. Necessary is examine whole person (not only speech)

Classification of logopedic diagnostics methods:

- observation method
- explorative method (conversation, verbal testing, written testing, practical testing)
- case study (study of results of phoniatric, ENT and neurologic examination, study of school materials)
- results analysis in diagnostics of SVPU
- instrumental methods – different indicators in case of incorrect pronunciation

Principles of diagnostics:

1. principle of complexity
2. principle of objective assessment
3. principle of quantification
4. principle of team approach
5. prinicple of continuum

Complex examination

1. contact establishing (children often don´t want to communicate, so draw a picture, tell a tale, …)
2. history – personal, family, social. Family – diseases of speech organs, accent, hearing, voice, left-handing). Personal – course of pregnancy, birth, development of child in early childhood (first words, sentences, injuries, …)
3. hearing examination – are made by phoniatrist, ENT specialist
4. examination of phonematic differentiation
5. examination of speech understanding
6. examination of speech production
7. examination of motoric functions
8. examination of laterality – made by psychologist
9. examination of social factors – relation ibetween child and family, friends in school, ….

Disorders of articulation

Dysartria

Dysartria is disorder of motoric realization of speech based on organic damage of CNS. It includes many types or syndromes, which are caused by difficulties in muscular control of speech mechanisms and are classified as a motoric speech disorders. There are variably present disorders of respiration, resonance, prosodia and phonation.

Etiology: damage of CNS can have different causes and can afflict different levels (from cortical to peripheral CNS). It determinates form and grade of dysartria. Prenatal causes are: prematurely born, infection of mother in pregnancy, congenital defects of motoric structures of CNS. Perinatal cases are e.g. damage of motoric areas by bleeding, asphyxia. Postnatal cases form the biggest etiological group – encephalitis, meningitis, intoxication, disease of vessels, inflammations of brain, head injury, degenerative diseases of CNS, brain tumors)

Symptomatology: symptoms depend on dysartria type, which dpends on localization and size of lesion. Most widely known is Kimmel´s classification of dysartria on:

- cortical (changed quality of voice, pressed phonation, slurred articulation, spastic character of speech, repeating of first syllables)
- pyramidal (spastic palsy of speech organs muscles, shallow breathing, rhinolalia aperta, slow, monotonic and barely understandable speech, prosodia defect)
- extrapyramidal (hypertonic-hypokinetic form – respiration defects, rigid speech, slurred articulation, rhinolalia aperta, repeating of syllables or words, changed prosodia; hypotonic-hyperkinetic form – afflicted breathing, change of voice, some speech sounds are accented, pace, dynamics and melody of speech is changed)
- cerebellar (difficult voice formation, speech is yelled with hesitations, often unintelligible)
- bulbar (type of peripheral palsy, afflicted muscles are atrophic, pronunciation of some speech sounds is afflicted, voice is dysphonic or even aphonic, rhinolalia aperta, monotonic speech – “hot potato speech”, sometimes is afflicted chewing and swallowing)
- mixed

Diagnostics: history and logopedic examination with description of mentioned symptoms. Detailed examination of articulation and speech-forming organs by ENT specialis (atonia of musces or paradox movements), including description of vocal cords mobility in stroboscopic light. Neurologic examination. Imaging methods of brain (especially MRI, but even PET or fMRI).

Dyslalia

Is the most widespread disorder of communication ability. It is inability to use single speech sounds or groups of them according to lingual-orthoepic standards. Afflicted is especially phonetic-phonologic level. Dyslalia develops during pronunciation evolvement. We differentiate 3 types: physiologic dyslalia (around 3 years of age), prolonged physiologic dyslalia and pathologic dyslalia (after 7.year).

Etiology: causes can be organic or functional. Functional dyslalia can be motoric (result of global lack of skill) and sesoric (insufficiently evolved motional and auditory differentiation). Organic dyslalia is

based on disorders of afferent or efferent nerve tracks, auditory tracks or CNS. Cause of dyslalia can be also interference between structure and function of speech and auditory mechanisms of brain. Causes can be differed to inner (physical immaturity, motoric discoordination, hearing disorders, insufficient sound discrimination, anatomical defects of speech organs, cognitive-linguistic imperfection, neuromotoric disorders) and outer (psychosocial influences). It is considered influence of heredity, emotional deprivation, improper speech pattern.

Symptoms: missing speech sounds (elimination, mogilalia), replacing speech sound with others articulatorly close (substituition, paralalia), impropaer pronunciation (distorsion, named according to afflicted speech sound: r – rotacismus, l – lambdacismus, s – sigmatismus, etc.).

Diagnostics: logopedic examination. Exclusion of other causes by ENT specialist. Other examinations in specific disorders.

Therapy: dyslalia is usually found in children and usually in that time it is corrected. Brain of child is permanently evolving (in 3.zear has 75% of its size), while in adults is fully evolved. That's why is harder to improve pronunciation in adults then in children. In addition, patient is used to his own pronunciation and the correct pronunciation is considered as strange. He hasn´t to meet negative attitude against his speech in his whole life.

Palatolalia

It is speech disorder, which accompany palatoschisis or palatocheiloschisis. It is result of organic defect, especially palatopharyngeal closure. Speech is developing on wrong base.

Etiology: term cleft defect doesn´t mean any fissure, but it means no connection of mesenchymal processes, which form base of bone and soft parts of face. Main mechanism of fissure development is later horizontalization of palatal plates (which are developing vertically at first) according to growth of whole face. Causes are multifactorial, there is combination of endogenous factors (heredity in 40%) and exogenous ones. Exogenous factors are teratogenic factors in first trimester of pregnancy (viral, parasite or bacterial infections), chemical substances (some mediaction, drugs), hypo or hypervitaminosis, physical factors (x-ray, irradiation, mechanic damage), wrong way of living of mother, her higher age (over 38 years), diabetes, metabolic defects, psychic trauma. Fissures are parts of variable syndromes (Apert´s, Crouson´s, Treacher-Collins´s, Sedláčkové sy)

Classification according to Burian:

Typical fissures:

I. group
   a. cheiloschisis (right, left, bilateral, total, subtotal) – fissure of lip
   b. cheilognathoschisis (goes through lip and premaxilla to the foramen incisivum) – fissure of lip and maxilla
   c. cheilognathopalatoschisis (right, left, bilateral) from foramen incisivum to uvula and similarly as in b.

II. group
    a. fissure of palate – isolated fissure of soft palate – staphyloschisis, fissure of uvula – uvula bifida, fissure of soft and hard palate – palatostaphyloschisis
    b. submucous fissure – isn´t visible, because mucosa is intact, afflicted is muscles or sometimes bone
    c. congenital short palate

Atypical fissures

- middle fissure of lip, fissure of lower lip
- macrostomia (lateral fissure of mouth isolated or combined with auricle deformations, transversal fissure of face)
- oblique fissure of face
- coloboma of eyelids
- fissure microforms (hereditary orofacial anomalies)

Classification according to Kernhan and Stark is used as well. This classification divides fissures according to position in relation with foramen incisivum on: fissures of primary, secondary or both palates and rare fissures.

Incidence of fissures of primary and secondary palate is in Czech Republic 1:530 (Škodová, Jedlička 2003) or 1:500-700 (Kerekrétiová 2000) of all life-born children. In submucous fissures is it 1:1200, in fissures of uvula is it 1:80 (Vitásková 2005).

Symptoms: main signs are anomalies of orofacial system, which depend on type of fissure, nose deformities, anomalies of teeth and jaws, velopharyngeal insufficiency, oronasal communication, hearing disorders (up to 90%). In some cases is afflicted suction and swallowing, which endanger the newborn by bad nutrition.

It is present speech disorder – palatophonia – which is term for three components: rhinolalia, resonant changes and hypekinetic dysphonia. Fissures even lead to problems in nonverbal communication (deformities in face limits mimics, analogous movement of mimic muscles can cause confusion, hypomimia in congenital short palate, own gesticulation system as a compensation in verbal section leads to loss of semantic communication channel, scars camouflage (moustache, make-up) change transfer of mimic information.

Most important specific disorder of resonance and articulation is from logopedic aspect - palatolalia. Different patients have different symptoms – according to extent and type of fissure, personality, timing and quality of treatment). There is affliction of vocals (typical hypernasal character) and consonants (significant changes). In some cases are missing consonants. Most frequent is dyslalia (39,6%), the least frequent is dental articulation (6,3%). Usually it is mixed form (in addition with palatal and compensatory articulation). Cleft defects are often connected with late speech evolvement (almost 50%), it hasn´t to be caused by velopharyngeal insufficiency, but it is rather combination of symptoms. Patolalia symptoms include compensatory mechanisms actively forming by individual (especially formation of linguopalatal or other closure as a replacement for nonfunctional velopharyngeal closure and passive, will independent mechanisms (hypertrophy of adenoids and tonsils, enlargement of Passavant´s wall).

Diagnostics: same methods as for hypernasality. Diagnosis is determined by logopedist, ENT specialist (baseic ENT examination, hearing examination, endoscopy of nose and velopharyngeal closure), stomathologist-orthodontist and other specialists. Improtant are imaging methods: static

(variable X-ray projections of skull, especially CT of skull, possibly MRI of velopharyngeal closure) and dynamic (position of articulation organs – especially velopharyngeal closure – and description of swallowing act).

Therapy: as team as diagnostics:

- surgical – plastic operations of palate, nose and lip. ENT operations – adenotomy, tonsillectomy, tonsillotomy, myringotomy with insertion of ventilation tubes, septoplasty. Maxillofacial surgery – bone grafts into interrupted alveolar arches, maxilla lengthening, etc. Stomathologic and orthodontic treatment
- internal – neonatological, pediatric and anaesthesiologic treatment
- rehabilitation – logopedic (speech), and phoniatric (voice and velopharyngeal closure)

Fissures have very broad issue. They are not only anatomic anomalies but even disorder of communication ability. Fissure can deeply influence whole personality of individual and cause great psychic problems. Thanks to rapid progress of medicine (especially plastic surgery and orthodontia) the situation is getting better, because with early intervention results can be very satisfying.

Disorders of voice sound

Rhinolalia/rhinophonia

We have two terms – rhinolalia (used by logopedists) and rhinophonia (used by doctors) – which are synonyms.

Rhinolalia is caused by disorder of nose resonance, which can be pathologically increased or decreased. This is caused by velopharyngeal insufficiency. Balance between orality and nasality is broken and resulting acoustic impression is speech “through nose” (hypernasality) or nasal resonance is not present (hyponasality).

Hypernasality

Pathologically increased nasality, or rhinolalia aperta, hyperrhinolalia. Nose and oral cavity aren´t connected when speech sounds are forming and articulation stream goes towards mouth instead towards nose. Nasal resonance participates in nasal speech sounds formation, and even in oral ones, which causes disruption of speech sound.

Etiology:

- Orality decerase (hypoplazia of palatum velum, elevation and retraction of dorsum linguae, small jaw angle).
- Velopharyngeal insufficiency
- Oronasal communication

Cause of hypernasality can be functional or organic – congenital and gained.

Organic congenital causes:

- Paresis (peripheral or central)
- Fissures of hard and soft palate including submucous ones

- Congenitally shortened soft palate
- Congenital defects (Sedláčkové sy)
- Insufficient or irregular development or pathological function of soft palate muscles
- Big epipharyngeal space – deep megapharynx

Organic gained causes:

- Paresis (peripheral or central)
- Perforation of soft or hard palate causing tissue atrophy
- Palate defects after surgery in oral cavity
- Velopharyngeal insufficiency
- Infections (flu, encephalitis, TBC, syphilis, diphtheria)
- Tumors
- Neurologic diseases (Parkinson´s disease, myasthenia gravis)
- Pseudobulbar paresis

Hypernasality caused by functional causes have irregular symptoms and intact function of velopharyngeal closure. Causes can be:

- Mental retardation
- Hearing disorder
- Psychogenic causes (hysteria, neurosis)
- Result of speech pattern simulation
- Habitual (as custom) rhinolalia aperta after adenotomy or tonsillectomy
- Muscular hypotonia causing velopharyngeal insufficiency
- Imitation
- Affectionate or negligent speech style

Hyponasality

Rhinolalia clausa, hyporhinolalia, is pathologically decreased or missing nasality.

Etiology: Hyponasality develops, if nasal expiratory flow cannot pass through nose due to closed velopharyngeal closure (or other obstruction). Articulation air stream goes through oral cavity and loses its nasality. Nasal speech sounds m, n, ň are heard as b, d, ď. Impassability of nasal cavity and/or nasopharynx can be partial or total, which causes denasality – absolute absence of nasality. Causes can be congenital or gained, organic or functional, temporary or permanent.

Hyponasality can be classified to anterior and posterior. Organic causes of anterior rhinolalia clausa are chronic or allergic rhinitis (inflammations of nasal mucosa), swelling of nasal mucosa in acute rhinitis, polyps, deformation of nasal septum, congenitally narrow nostrils, tumors, orofacial deformities, congenital syndromes. Problem is impassability of nasal cavity. If nasopharyngeal cavity is impassable, it is called posterior rhinolalia clausa. Impassability can be caused by hypertrophy of adenoids or tonsils, syphilis, atresia, tumors, bulky Passavant´s wall, deformation of nasal septum after injuries, etc. Functional causes are functional muscles disorders of soft palate causing increased activity and strength of velopharyngeal closure.

Hyper-hyporhinonasality – rhinolalia mixta. It happens in presence of velopharyngeal insufficiency and impassability of nasal or nasopharyngeal cavity. Causes can be organic or functional. Afflicted are not only vocals and sibilants, but even nasal consonants.

Diagnostics: basic examination tests:

- Gutzman´s A-I test
- Czermak´s test
- Test of face inflation
- Test with otophone

Instrumental examination techniques: velopharyngometer, manometer, spirometer, ultrasonography, cephalometric examination, videofluoroscopy, endoscopy, fibroscopy, X-ray, EMG, electropalatography, articulography, sonographic and aerodynamic analysis.

Therapy: cooperation of doctor and logopedist. In hyponasality logopedist aims at nose breathing training and correct articulation of nasal speech sounds. In hypernasality is used training for function improvement of velofaryngeal closure and thanks to it better resonance. Training can be active (blowing, whistling, suction, gargling, yawning, swallowing, etc.) or passive (activation of diaphragm and soft palate by variable activities – massage, electrostimulation, gag reflex, correct posture, etc.). In mixed form is therapy aimed on dominant component.

Rhinolalia can develop in children and in adults. Surgery in children doesn´t necessarily mean problems regression. Usually is needed long term reeducation with training according to type of rhinolalia. In adults can rhinolalia sometimes cause only glimpse of persistent mimic disorders as a residue of compensatory mechanisms in limited possibilities of articulation.

Disorders of speech fluency

There is tumultus sermonis (babbling) and balbuties (stammer). Both those disorders were considered as speech neurosis. Now it is thought, that they have organic cause. Big problem is relatively frequent combination of both. In this case we have to insist on differential diagnostics.

Tumulus sermonis

Definition: communication ability disorder. Person is not aware of it, it has limited range of attention, have afflicted perception, articulation and speech formation. There is also disorder of mind processes based on heredity. Tumulus can be caused by central speech disorders and influences all communication ways: reading, writing, rhythm, musicality, behavior. It is disorder of speech fluency characterized by extremely fast rate of verbal production.

Tumulus isn´t isolated nosologic unit, it evokes syndrome. It is nonspecific variable disorder. People with this disorder have speeded up not only speech but even all behavior, we can say - whole personality.

Symptoms: overbrash rate of speech and afflicted articulation (person cannot maintain correct pronunciation), repeating and reduction of syllables, respiratory dysrytmia, voice disorders (develops due to uncoordinated respiration and phonation), dysprosodia (afflicted melody – often monotonic, rhythm, dynamic accent).

Speech is afflicted even from linguistic aspect, because thanks to thinking disorganization (thinking are faster than speech or vice versa) are formulated linguistically incorrect sentences, speech is sometimes without content and have poor syntax. Semantic non-fluency merge into grammatical and after that to phonetic non-fluency. Person is not aware of it, because it insufficiently controls its answers. We can see even nonverbal manifestation – movement, walking, global motorics are accelerated too. Sleeping disorders can appear. It can influence even writing, which can be disintegrated and dysgraphical.

Etiology: not clearly known.

Balbuties

it is the most serious and most apparent disorder of communication ability. It is solved by separated section of logopedy (balbutology). In ….. is afflicted rate of speech, in balbuties is afflicted speech dynamics. Primary it is disorder of non-symbolic processes (fluency disorder), which afflict even symbolic processes (dirupting effect of balbuties on speech formulation, avoiding critical words by their paraphrase, in severe forms impossible speech understanding). It usually influences whole personality and lifestyle. We consider it a complex disorder of organs coordination participating in speaking. It is most apparently manifested by characteristic unwillingly fluency interruption. Symptoms can be seen on all ligual levels.

Etiology: supposed 3 etiologic groups:

- organic – dynamic disorder of feedback control of motoric mechanisms in ralization of speech in subcortical basal ganglia (heredity, metabolic disorders, vegetative lability, congenital speech weakness, afflicted auditory feedback, balbuties as a result of discoordination of brain hemispheres)
- developing primarily as a neurosis – negative influence of social environment (espeially long term neurotization), pschotrauma, simulation, avoiding behavior, disorder of maternal interaction
- secondary neurotic disorders based on organic predisposition – combination of both previous causes

Possible is mutual inosculation of single causes. It complicates the situation, because if we cannot recognize primary cause of disease, we cannot cure it causally but only symptomatically.

Symptoms: are divided into inner and outer.

Inner symptoms: in most severe cases fear from speech and avoiding direct verbal contact or even refusal of social life (logophobia).

Outer symptoms:

- respiration disorder (interrupted, irregular breathing, disorder of breating moves or inspiratory voice formation)

- phonation disorder (hard voice starts or dysphonia)
- articulation disorder
- increased phonation pressure and resulting longlasting speech spasm
- prosodic factors disorders (monotonic speech, fluctuating rate)
- embolophrasia (so called vocal emboli: words or speech sounds – e.g. hm, so, yes – patient use them to overcome the speech spasm)
- paraphrase or synonyms
- co-verbal behavior disorder (grimaces, dithering, winking, motoric agitation, swallowing, co-movements of upeer and lower limbs, sometimes even whole trunk, Froschels´s symptom, etc.)
- somatic symptoms (increased blood pressure and muscular tension, metbolism disorders, insomnia)
- other symptoms – difficulties in writing, reading, disorder of smooth motoric

All symptoms are influenced by situation of afflicted person and by his psychic and physic state.
Forms of balbuties:

- clonic
- tonic
- mixed

AS we can see above, people with this disorder are in difficult situation. In adults it is usually chronic balbuties, characterized by dysfluencies, increased effort during speech, psychic tension, and often even logophobia
Diagnostics: logopedic examination, psychologic examination, neurologic examination (EMG), ENT examination (acoustic feedback test – Lee efect)
Therapy: synthesis of various logopedic and psychologic approaches. Pharmacologic treatment doesn´t have longlasting effect.

Disorders of speech development

Speech development is complicated procedure, which is influenced by many inner (congenital defects, hearing and sight development, motoric and intelect developmetn, etc.) and outer factors (environment, education, speech model). If some of the compounds isn´t all right, speech development disorder results which negatively influences psychic development of child, it´s school outcomes, personality formation, hobbies and future profession
Speech development disorder is structural and systemic disorder of one or more areas of speech development (acquiring native language, development of language abilities) according to age of child. We can see defects in all language areas. Term “evolutionary taciturnity” consists of two diagnoses – simple delayed speech development and evolutionary dysphasia.

Simple delayed speech development

It is not classified as a development speech disorder, because speech is not structurally disrupted, speech development is only delayed in one or more language areas. However, it is important to be aware of this disorder, because it can be the first signal of real development speech disorder.

Evolutionary dysphasia

It is specific disorder of speech development which causes difficulty or inability to learn verbal communication, when conditions for speech development are adequate.

Etiology: multifactorial. It is supposed, that it is caused by bilateral diffuse cortical lesion in speech areas of brain in early development. Heredity and gender also play role in development of psychic functions lateralization in brain hemispheres. It exists so called congenital speech weakness as well. Coincidence was found between brain development disorder and specific speech development disorder. Even environment has its role in this. Cause can be divided into genetic, congenital and gained.

Symptomatology: is very extended. Most significant disorder is present in speech and language. It means e.g. small word-stock, short memory disorders, inability to keep plot line, disorder of sound signals timing, false speech sound order, dysprosodia, dysrythmia and dysmusia. This disorder can usually cause specific evolutionary learning defects in scholl age. We can see also symptoms of disorder based on neurologic, audiologic and phoniatric examination, strange behavior (disorders of activity and awareness, fatigue, impulsivity, disorder of right-left orientation, disproportion between verbal and nonverbal part of intellect, colour sense disorder, etc.), motoric defects (fine and rough motorics and oromotorics), disorder of tactile sensibility.

Diagnostics:

- neurologic examination – basic examination, EEG, cognitive evoked potentials
- logopedic examination
- audiologic examination – examination of hearing, central hearing disorders and vocal organs
- psychologic examination – examination of intellect (verbal and executive parts), laterality, concentration, awareness, memory, etc.

Specific development speech disorder is very wide and serious problem. We put an accent on complex approach, differential diagnostics and cooperation with school and child´s family. In adults it manifestates as a specific learning disorder.

Aphasia

Aphasia is total or partial inability to receive and send symbolic codes of spoken or written speech – loss of already gained communication ability. It is a lingvistic , neurogennous and cognitive disorder. It is part of group of higher cortical function disorders.

Etiology: it develops on the base of organic lesion of brain. Most common causes are strokes (haemorrhagic or ischemic), injuries of brain (commotio cerebri, brain contusion, brain tumors, inflammations, degenerative diseases of central nervous system (Alzheimer´s disease, Pick´s disease), intoxication (by drugs or toxic gases). Special cathegory is children aphasia (infantile), which is caused by sudden interruption of normally evolving central nervous system (CNS). Causes are similar (excluding strokes – they are not so frequent as in adults). Most common is craniocerebral injury (Čecháčková, 2003).

Symptomatology: symptoms of aphasia depend on type of CNS damage, where it is localized and depends on age of patient as well. Symptoms are present in variable quality, quantity and combinations. Modern aphasiology describes those symptoms:

- fluency disorders
- paraphasia (deformation of words of varible grade and type). They are divided into paraphasia phonemic (deformed but understandable word), jargonic (deformed and inunderstandable word) and semantic (word replacement with another word of similar meaning)
- paraphrasia (disability or even inability to make sentences, only fragment remains – only one word, so called word wreckage.
- Perseverance (using previous signal in other answers, even if th signal is not in effect)
- Logorrhea (very fast speech with decreased intelligibility)
- Anomia (desription disorder – man cannot find adequate word for specific thing)
- Neologisms (new word with no sense)
- Understanding disorders (very frequent, so we must examine this)

In aphasia there can be afflicted all speech modalities – receptive and expresive, speech spoken and written.

There are many different classification (Boston, Kiml, Lurij, Hrbka).

According to Olomouc classification is brain lesion localized in specific lobe characterized by specific symptoms, which form specific cortical disorders. We can divide aphasia into:

- **expresive aphasia** (paraphrasia, paraphasia, non-fluent speech, understanding to spoken speech is normal)
- **integration aphasia** (main characteristics is so called Gerstmann´s syndrome – agnosia of fingers, right-left desorientation, dissociated agrafia, dyscalculia)
- **perception aphasia** (disorder of speech understanding and decoding, often logorrhea, ability to produce speech is saved, but speech is usually unintelligible. In case of severe affliction man don´t understand even its own words, neologisms, perseverance, graphia, lexia and drawing disorder)
- **amnestic aphasia** (least severe form – common latencies in speech, anomia, semantic paraphasia, understanding isn´t afflicted)
- **global aphasia** (most severe form, it afflicts all cortical functions – patient cannot understand to spoken speech, cannot speak, global desorientation)

Diagnostics, therapy and rehabilitation are interdisciplinary matter. We try to reach optimal communication level according to level of brain damage.

Diagnostics: every aphasiologic school has its own approach to diagnostics. However, all concentrate on examination and assessement of spontaneous speech, understanding, repeating and description, written speech as well as spoken. There are many testing batteries. We use modification of Token test, Lyrij´s neuropsychologic examination.

Therapy: neurologic tratment according to etiology, logopedic rehabilitation of speech.